RHEUMATOLOGIC AND IMMUNE-MEDIATED DISEASES

[Abstract:0003]

DILATED CARDIOMYOPATHY AND ANTI-TNI ANTIBODIES IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Aim: Dilated cardiomyopathy DCM) is a rare occurrence in systemic lupus erythematosus (SLE). Anti-Troponin I (anti-TnI) antibodies were found in 12% of a lupus cohort and were not present in patients with other rheumatic diseases. This raises the possibility of the antibodies being associated with cardiac involvement.

Methods: Twenty-three lupus patients (diagnosed according to the ACR criteria) were positive for anti-TnI antibodies and they were matched according to age, gender and ethnicity by anti-TnI negative lupus patients. All patients had 2-D echocardiography performed. Echocardiographic features of DCM were: (a) left ventricular (LV) dilation, (b) systolic dysfunction with impaired global contractility, (c) normal LV wall thickness, and (d) LV diastolic dysfunction with elevation in LV filling pressure. Lupus-related autoantibodies profiles were obtained through chart review. The odds ratio and Fisher's exact test were calculated.

Findings: The clinical and autoantibodies profiles were comparable in both groups. The age range of the study and control groups were 21 years to 64 years, with the mean age of 38.6 years and 39.7 years respectively. Two patients (1 male and 1 female) from the anti-TnI positive group fulfilled echocardiographic features of DCM while no case was detected in the anti-TnI negative group. The odds ratio is infinity and the Fisher's exact test (2-tailed) showed a p value of 0.487

Conclusions: The finding of dilated cardiomyopathy in the anti-TnI positive group is interesting though statistically not significant due to the small numbers studied. Study of a larger cohort may clarify the observed finding.

Keywords: *anti-TnI antibody, cardiomyopathy, lupus*

[Abstract:0010]

MALE SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH ACHALASIA: A RARE CASE

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Systemic lupus erythematosus (SLE) is a complex autoimmune disease with multisystem involvement including gastrointestinal involvement in various forms. In some rare cases achalasia may present. In this case report, we aimed to discuss the process of diagnosing systemic lupus erythematosus in a male patient who presented with dysphagia, anemia and weakness.

A 53-year-old patient was diagnosed with achalasia three months ago due to dysphagia, but the etiology could not be found. The patient has admitted to our clinic with complaints of weakness, rashes, dysphagia, joint pain and weight loss. Initial tests revealed coombs positive hemolytic anemia, hypocomplementemia, elevated sedimentation levels. Immunological parameters were positive for systemic lupus heritematosus. Cranial magnetic resonance imaging (MRI) revealed chronic ischemic changes. Patient was treated with pulse steroid followed by oral steroid and six cycle of cyclophosphomid. The patient is currently in remission and is being monitored with mycophenolate mofetil and aspirin treatments. Achalasia is a rare condition characterized by aperistalsis of the esophageal body and failure of the lower esophageal sphincter to relax. In a prospective study, it was observed 9% of patients who underwent high resolution manometry due to dysphagia had underlying rheumatological diseases.

The most common were systemic sclerosis (2.7%), followed by rheumatoid arthritis (2%) and systemic lupus erythematosus (1%). In our patient the presence of dysphagia symptoms, anemia and complement deficiency led us to consider this preliminary diagnosis. The fact that patient had autoimmune hemolytic anemia, involvement in cranial MRI and positive anti-dsDNA may indicate the potential for more severe complications in the future.

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Therefore, in patients presenting with achalasia and anemia, even in male patients, systemic lupus erythematosus should be considered.

Keywords: *achalasia, dysphagia, lupus*

[Abstract:0054]

BEYOND DRYNESS

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Case Description: A 23-year-old patient, without important medical history, was admitted for 3 days with asthenia, fever and odynophagia. 48 hours later, he presented a generalised exanthema with severe oral mucosal and conjunctival involvement. He denied recent travel and transfusions. Tortuous course with conjunctival adhesion, and oral mucosal bleeding with anaemia and inability to swallow.

Clinical Hypothesis: Erythema multiforme.

Diagnostic Pathways: Skin biopsy confirmed suspected erythema multiforme. Cultures and serology were negative. Autoimmunity: ANA positive (1/320) mottled pattern, anti-SSA/Ro >640, and anti-SSB/La >640 antibodies. After a positive autoimmunity, a parotid scintigraphy was performed and primary Sjogren's syndrome (pSS) was confirmed. Subsequently, the patient presented numerous associated complications:

- Corneal perforation and abscess of the left eye. 2 corneal transplants and, finally, a left eye prosthesis.
- Apical bullae together with diffuse cylindrical bronchiectasis which have led to repeated respiratory infections and pneumothorax on several occasions requiring surgical resection.
- Chronic renal disease without evidence of glomerulopathy with metabolic acidaemia due to distal renal tubular acidosis secondary to interstitial nephritis.

Discussion and Learning Points: pSS is one of the most common chronic autoimmune diseases, characterised by an inflammatory infiltrate affecting exocrine glands, giving rise to the characteristic "dry syndrome" behind which a systemic phenotype of highly variable prevalence and sometimes occurring prior to the diagnosis of pSS can hide. Beyond dryness and its apparent benignity, pSS can have severe extraglandular involvement. These should be actively monitored and detected. Early diagnosis and treatment will determine chronic damage in these patients.

Keywords: *primary Sjogren syndrome, erythema multiforme, extraglandular involvement*



Figure 1. *Mucosal lesions.*

[Abstract:0086]

ACUTE CARDIAC TAMPONADE AS ELDERLY SYSTEMIC LUPUS ERYTHEMATOSUS DEBUT

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Case Report: a 75-year-old woman presented with asthenia, fever, hypotension and syncopes. She had a medical history of hypertension, anticoagulated paroxysmal atrial fibrillation, without structural heart disease, and chronic pain with rheumatoid positive factor diagnosed as adult rheumatoid arthritis.

Physical examination revealed hypotension, normal oxygen basal level, no fever, irregular heart sounds at more than 100 bpm, no extratones, lung hypoventilation, and erythematous lesions on the back.

On urgent blood test we found hyponatremia, NT-proBNP 9531 pg/mL, normal procalcitonin levels, C-reactive protein 109 mg/L, bicitopenia: 9.2 g/dL hemoglobin, 670 g/dL absolute lymphocytes, 596 g/dl platelets, prothrombin time 15.6 seconds (62%). Chest X Ray showed cardiomegaly and blunting of the costophrenic angles. Electrocardiogram showed atrial fibrillation at 130 bpm, with low voltages. Urgent transesophageal echocardiogram was done and demonstrated severe pericardial effusion with cardiac tamponade.

After anticoagulation reversal, pericardial diagnostic and therapeutic drainage was done. Secondary pneumopericardium appeared. Main result to emphasize were: positive ANA high titles and ANti-DNA.

Afterwards routine analysis showed ANAS an anti-DNA positive and positive SSA/Ro-52, 60 (DOT), SSB/La (DOT), Ac anti nucleosoma. Systemic lupus erythematosus (SLE) in the elderly, appears after 50-60 years old, with clinical manifestations that differ from classic SLE. In this patient the possibilities of infectious or neoplastic origin were dismissed using image and laboratory diagnostic tests. Presence and positiveness of ANA and ANti-DNA on pericardial effusion are specific markers of SLE.

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Keywords: *systemic lupus erythematosus (SLE), elderly, cardiac tamponade, pericardial effusion.*

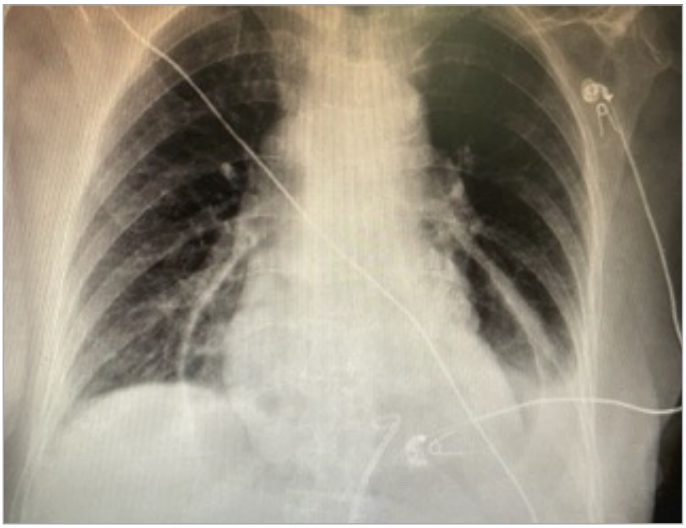


Figure 1. *Pneumopericardium after Pericardial Drainage.*



Figure 2. *Previous Chest Xray.*



Figure 3. *Urgency Chest Xray showing Cardiomegaly.*

[Abstract:0095]

NEUROLOGICAL INVOLVEMENT IN CONNECTIVE TISSUE DISEASES: AN EXPERIENCE FROM AN INTERNAL MEDICINE DEPARTMENT

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Introduction: Neurological involvement (NI), whether central or peripheral, in connective tissue diseases (CTD) is rare. It is likely underdiagnosed. It can be severe, potentially life-threatening or affecting the functional prognosis.

Aim: To study the clinical and paraclinical profile, treatment and follow up of NI in CTD.

Materials and Methods: A retrospective, descriptive, observational study was conducted, over a period of 55 months, involving 67 patients with CTD, mean age was 37.5 years. Sex ratio was 8.5. The diagnosis was established based on clinical, immunological, radiological, and histological criteria.

Results: Eleven patients with CDT and NI were included in the study. Mean age was 35.9 years, sex ratio was 10. The main symptoms observed were paresis, paresthesia, quadriplegia, hemiplegia, loss of balance, and myalgia. The onset of symptoms was acute, subacute, or progressive. NI led to the diagnosis in all patients. It was isolated or associated with other clinical symptoms. It was central but most often peripheral. Specific treatment was administered to all patients, including high-dose corticosteroids+/-immunoglobulins+/-immunosuppressive therapy. The short- and medium-term outcomes were favorable, with partial improvement of clinical symptoms in all cases and long-term neurological sequelae persistence in two cases.

Conclusions: Neurological involvement in connective tissue diseases is rare. It can be isolated and revealed the disease, representing a real diagnosis challenge. It most often involves motor impairment ranging from weakness to complete paralysis, with or without paresthesia. It would most often be of peripheral origin (at least based on our sample). Early specific treatment and good adherence to background treatment ensure a favorable outcome without sequelae.

Keywords: *connective tissue diseases, neurological involvement, central, peripheral, motor, paresthesia*

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[Abstract:0100]

UNVEILING CLIPPERS SYNDROME: A CASE PRESENTATION

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A 58-year-old male was admitted to the Emergency Department with a recent-onset lower back pain and weakness in his lower limbs that had been evolving over several months. Notably, he was a heavy smoker with a 40-pack-year history but had no known prior medical conditions.

Imaging studies, revealed multiple osteolytic lesions with pathological fractures at L1, L3, and L4, and a liver biopsy confirmed extensive small-cell carcinoma without an identified primary site.

After receiving radiation therapy for the vertebral lesions, the patient initially showed clinical improvement. However, prior to commencing systemic treatment (Carboplatin-Etoposide), he experienced subacute neurological symptoms, including moderate dysarthria, bradyphrenia, behavioral alterations, and a previously unknown gait disturbance.

With suspicion of CNS tumor infiltration, a cranial MRI was conducted, shown in Figures 1, 2 and 3. These findings were highly suggestive of CLIPPERS (Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids).

Given the patient's vertebral fractures, a lumbar puncture was not feasible, but a exhaustive study was completed with all the results returning negative.

In light of the CLIPPERS suspicion, corticosteroid therapy was initiated (1 mg/kg), leading to significant clinical improvement and the immediate resolution of neurological symptoms. Following two weeks of clinical stability on tapering corticosteroids, systemic treatment commenced without incident.

In summary, CLIPPERS syndrome, an exceedingly rare and enigmatic condition, presents significant diagnostic challenges. This case emphasizes the crucial need to consider CLIPPERS in patients with subacute neurological symptoms, even without definitive cerebrospinal fluid analysis, as it responded favorably to corticosteroid treatment.

Keywords: *CLIPPERS syndrome, paraneoplasic syndrome, inmune-mediated*

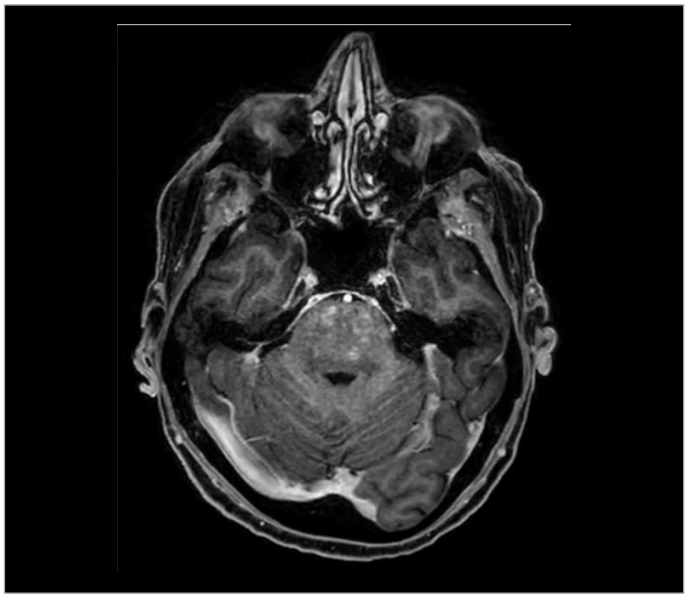


Figure 1. *T1 + Contrast (Axial): Following the contrast administration, multiple patchy enhancing areas are observed in the pons.*

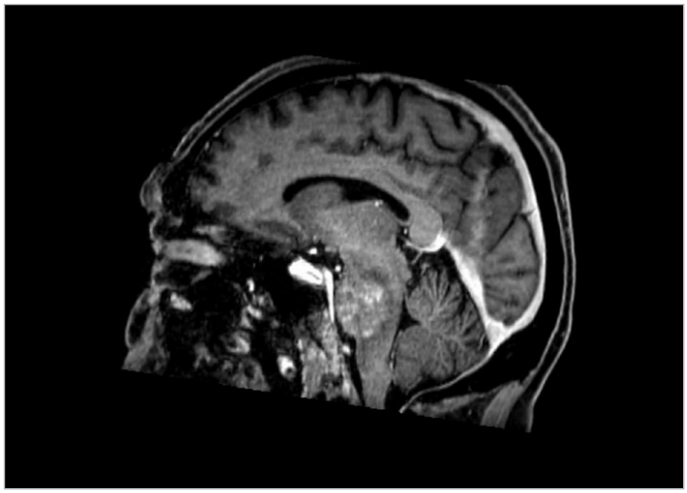


Figure 2. *T1 + Contrast (Sagittal): After contrast administration, multiple patchy enhancing areas are observed, predominantly in the pons.*

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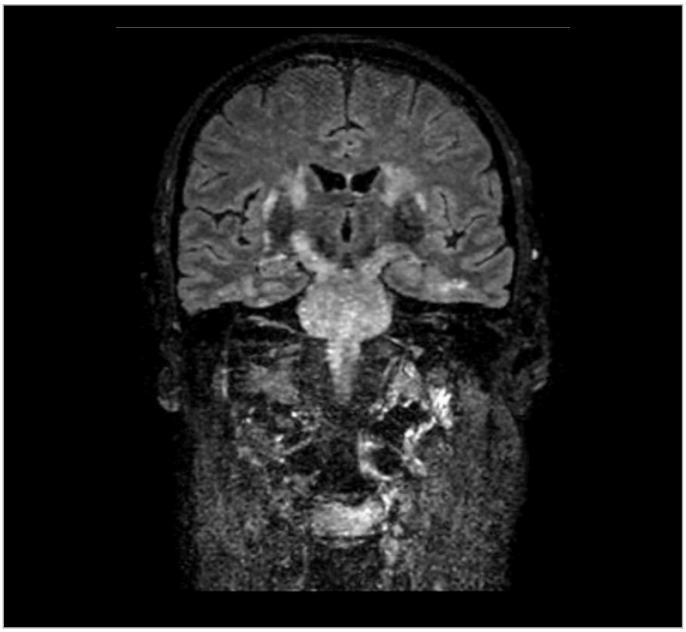


Figure 3. *FLAIR (Coronal): Predominant signal hyperintensity in the pons, but also evident in the medulla oblongata, both brainstem peduncles, external capsules, and internal capsules.*

[Abstract:0137]

ATYPICAL SHU WITH TROMBOTIC MICROANGIOPATHIA SECONDARY TO ENDOVESICAL BCG INSTILLATION

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65-year-old male with high-grade bladder carcinoma with TUR and periodic BCG instillations. He came to the emergency department for febrile syndrome 3 hours after the last BCG instillation. Laboratory tests showed acute deterioration of renal function (creatinine 2.34 mg/dL), elevated acute phase reactants (CRP 193 mg/L, procalcitonin 63 ng/mL), leucocytes 11600 and urine systemic test with 500 leucocytes.

It was decided to start empirical antibiotic therapy with ceftriaxone 2g/24h and gentamicin 500mg/12 hours while awaiting cultures. The patient presented a torpid evolution during admission, with data of severe acute renal failure requiring haemodialysis, maintaining creatinine levels of 6g/dL, proteinuria of 1g without haematuria.

On the other hand, coagulopathy which, having ruled out other possible causes such as neoplastic progression or DIC, ADAMTS13 deficiency, is assumed based on the analytical data and as a diagnosis of exclusion, aHUS with secondary thrombotic microangiopathy probably related to BCG instillation. Treatment was started with eculizumab 900 mg weekly for 4 weeks with excellent clinical and analytical evolution.

aHUS is a disorder of the microvasculature, clinically defined by microangiopathic haemolytic anaemia (Coombs negative) and thrombocytopenia, usually involving the kidneys and manifesting with haematuria, oligoanuria and renal failure.

Secondary TMA is derived from an imbalance between immunity, coagulation and complement, altered by precipitating factors in patients predisposed to TMA by multiple genetic determinants. Given the relevance of complement as a producer of endothelial damage in the pathogenesis of both processes, it has been proposed to treat with Eculizumab, with excellent response.

Keywords: *BCG instillations, renal failure, eculizumab*

[Abstract:0170]

CHRONIC KIDNEY DISEASE DUE TO AA TYPE AMYLOIDOSIS IN A PATIENT WITH HIDRADENITIS SUPPURATIVA

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Aim: A 53-year-old man was found o have proteinuria and high creatinine levels (stated as 3.84 mg/dL while it had been 0.74 mg/dL one year ago) during his routine cardiology clinic visits and was diagnosed as chronic kidney disease due to AA type amyloidosis secondary to hidradenitis suppurativa.

This case describes an uncommon co-occurrence of AA amyloidosis, as one of the many causes of chronic kidney disease, and hidradenitis suppurativa.

Methods: Possible causes of acute kidney injury and laterly chronic kidney disease were gone over with biochemical tests, radiological and pathological methods.

Findings: Protein discharge in a 24-hour urine collection was concluded as 24376.5 mg. In search of any cause of nephrotic syndrome, amyloidosis was suspected due to his chronic inflammatory condition (Hidradenitis Suppurativa). A salivary gland biopsy from the inferior labium of the mouth showed amyloid accumulation. For both hidradenitis suppurativa and newly diagnosed amyloidosis, biological agents were the treatment of choice.

Results: Amyloidosis should be considered in the differential diagnosis of proteinuria, especially in cases of chronic inflammatory conditions. Furthermore, screening for proteinuria, as a manifestation of chronic inflammation in hidradenitis suppurativa, with urine dipstick tests can be beneficial not only for early detection of amyloidosis but also for decreasing the need of biological agent use.

Keywords: *hidradenitis, amyloidosis, kidney*

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Figure 1. *Active hidradenitis suppurativa lesion on the right gluteal region.*

The patient had scars of previous hidradenitis suppurativa lesions throughout her belly and bilateral lower extremities. On his right gluteal region, in an area of 8 cm in width, he had active lesions that are a combination of small lumps, sinus tracts and purulent discharge.

	Value	Reference
Hemoglobin	9.1 g/dL*	13.5-16.9 g/dL
MCV	86.3 fL	81.8-95.5 fL
Leukocyte	16.5 x 10^3/µL	3.91-10.9 x 10^3/µL
Thrombocyte	479 x 10^3/µL	166-308 x 10^3/µL
C-Reactive Protein	3.84 mg/dL*	0-0.8 mg/dL
Erythrocyte Sedimentation Rate	118 mm/hr*	0-20 mm/hr
Creatinine	2.89 mg/dL*	0.67-1.17 mg/dL
Estimated Glomerular Filtration Rate	23.71 mL/dk/1.73m²*	>60 mL/dk/1.73m²
Total Protein	4.79 g/dL*	6.4-8.3 g/dL
Albumin	1.67 g/dL*	3.5-5.2 g/dL

INR	1.17	0.8-1.2
Fibrinogen	900 mg/dL*	180-350 mg/dL
aPTT	38.8 seconds*	22.5-32 seconds
D-Dimer	1.13 mg/L*	0-0.55 mg/L
Urinary pH	5.5	5.5-7.0
Urine Specific Gravity	1.020	1.010-1.030
Urine protein	1617.5 mg/dL*	Negative
Protein discharge in a 24-hour urine collection	24376.5 mg*	<150 mg/day
Antinuclear Antibody	Negative	Negative
Antineutrophil Cytoplasmic Antibody	Negative	Negative
Complement 3	1.1 g/L	0.9-1.8 g/L
Complement 4	0.436 g/L	0.1-0.4 g/L
Immunoglobulin G	1100 mg/dL	751-1560 mg/dL
Immunoglobulin A	6.95 g/L*	0.7-4 g/L
Immunoglobulin M	81.9 mg/dL	46-304 mg/dL

Table 1. *Laboratory Data.*

[Abstract:0172]

LIVER DISEASE AND CRYOGLOBULINAEMIA

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Case Description: A 66-year-old man with a history of type 2 diabetes mellitus (DM) consulted for two months' evolution emetic syndrome and pruritus. Blood and urine tests showed renal failure with 3mg/dl creatinine, hypoproteinemia and non-selective 24 hours proteinuria of 4g. Serology for hepatitis C virus (HCV) was positive with high viral load. In addition, abdominal ultrasound showed non-cirrhotic chronic liver disease; renal doppler ultrasound was normal.

Clinical Hypothesis: As differential diagnosis of acute renal damage with nephrotic range proteinuria we considered prerenal origin with acute tubular necrosis already established in relation to vomiting, diabetic nephropathy and finally parenchymal cause, assessing its possible relationship with HCV.

Diagnostic Pathways: Prerenal and diabetic causes seemed less likely due to null response to volumen replacement and the history of short-standing DM with good glycemic control. Regarding parenchymal causes, an autoimmune profile was requested resulting antinuclear antibodies, rheumatoid factor and cryoglobulins negative, without evidence of complement consumption. Moreover, blood and urine immunoelectrophoresis was performed with no evidence of monoclonal peak.

Finally, considering the possible association of acute kidney injury and HCV related glomerulopathies, a renal biopsy was performed, revealing the existence of cryoglogulinemic glomerulonephritis type II related to HCV infection. The patient was submitted for antiviral treatment.

Discussion: Type II cryoglobulinemic syndrome is responsible for up to 80% HCV associated glomerulopathies. The absence of analytical data suggestive of it in our case, negative rheumatoid factor, lack of complement consumption and absence of serum cryoglobulins, are explained by the intermittent and fluctuating nature of it.

Keywords: *cryoglobulinanemia, hepatitis virus C, VHC*

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[Abstract:0205]

DRUG- INDUCED TROMBOCYTOPENIA AND ADVERSE REACTION TO RITUXIMAB: AN UNUSUAL CASE

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Introduction: Chemotherapy-induced thrombocytopenia is a common complication. Rituximab, an anti-CD20 monoclonal antibody, induces lysis of B cells only. We describe an acute thrombocytopenia induced by Rituximab associated with anaphylaxis and acute kidney failure.

Case Presentation: A 66-year-old woman affected by splenic marginal lymphoma underwent therapy with rituximab with premedication. During the first infusion, patient showed bilateral low back pain followed by widespread pricking pain, dyspnea and hypotension. She was treated with steroid, O_2, ephedrine and sent to the ED. Emergency blood tests revealed severe thrombocytopenia (15,000/mm^3), confirmed in citrate, and acute renal failure (creatinine: 1.78 mg/dL). The platelets at baseline were 130,000. Admitted to our department, on therapy with deltacortene 50 mg daily, there was a slow recovery of platelet values (25,000->35,000->43,000->60,000/mm^3) and a return of basal creatinine. Concomitant infections were excluded. Complement C3 was slightly reduced (85 mg/dL) 3 days after the event, with C4 within limits. The patient was discharged to continue hematological follow-up.

Conclusions: Rituximab-induced thrombocytopenia is a condition of rapid onset with unclear pathogenesis. In this case the association with an infusion reaction, similarly to what happens with IV immunoglobulins, suggests immunological mechanisms (complement-mediated or lysis) responsible for the reaction to the drug.

Keywords: *rituximab, trombocytopenia, adverse drug reaction*

[Abstract:0253]

CLASSIC PAN UNVEILED: A DIAGNOSTIC JOURNEY THROUGH PET IMAGING

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Case Report: A 79-year-old male patient was admitted to the hospital with a one-month history of fever, with a vespertine pattern, night sweats, fatigue, hyporexia, and weight loss of 2Kg. No abdominal pain, myalgias, arthralgias or other symptoms in the systems review was referred. He had hypertension, and a second antihypertensive medication was recently introduced.

Upon examination, the patient had a very good general condition, with an elevated blood pressure (170/100 mmHg). Normal temporal pulses and no lymphadenopathy was found. Regular rhythm without murmurs. No signs of peritoneal irritation or megalies. No skin lesions, edema, or signs of arthritis.

Laboratory tests revealed anemia normocytic (Hb 10.1mg/dl) with an elevated erythrocyte sedimentation rate (ESR: 80mm/h) and C-reactive protein (CRP: 8.9mg/dl). An autoimmune panel (including ANA, ANCA, Complement, RF/aPPCC, and immunoglobulins) and microbiological screening were conducted, yielding negative results. No alterations were found in the elemental and sediment analysis. A CT angiography scan was normal, and a temporal artery biopsy was performed, with negative diagnostic results.

A PET/CT was performed, described in Figure 1.

The patient was diagnosed with classic polyarteritis nodosa so a treatment with high-dose corticosteroids and immunosuppressive agents (Methotrexate) was initiated, with a disappearance of the symptoms and an excellent ambulatory evolution.

This case underscores the diagnostic challenges associated of a vasculitis due to the absence of specific symptoms or signs. PET/CT imaging can be employed during the initial stages of polyarteritis nodosa for an early diagnosis, particularly in the absence of vasculitis-related complications.

Keywords: *polyarteritis nodosa, vasculitis, PET/CT*

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Figure 1. *PET/TC maximal intensity projection (MIP) showed increased tracer uptake forming lines in leg muscle compartment that seems to paint the vascular three. Images also reveal high tracer uptake in bilateral humeral and internal mammary arteries. Intense bone narrow and slight splenic hypermetabolism was already disclosed. Images did not reveal increased uptake in aorta neither temporal arteries. These findings suggest vascular inflammation of small and medium size vessels, majority located intra or inter-muscular tissue in limbs. Bone narrow activation and hypersplenism demonstrate non-specific inflammation associated reaction.*

[Abstract:0269]

DESCRIPTIVE ANALYSIS OF PATIENTS WITH SARCOIDOSIS UNDER FOLLOW-UP BY THE AUTOIMMUNE DISEASES UNIT IN A THIRD LEVEL HOSPITAL

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Sarcoidosis is a systemic disease that predominantly affects women between 20-50 years of age. The most affected organs are the lungs, lymph nodes and skin. Diagnosis is based on histological demonstration of non-caseating granulomas in patients with clinical or radiological suspicion and corticosteroids are the first line of treatment.

We made a retrospective description of the clinical characteristics, diagnostic process and therapeutic management of 57 patients under follow-up for sarcoidosis in our center between January 2008 to May 2023.

The average age was 53±14 years (19-87), with a female predominance (70.2%). The most frequent clinical presentations were pulmonary and cutaneous, both with 43.9%. A second autoimmune disease was diagnosed in 5.3%, with diagnoses of Sjogren's syndrome (n=2) and psoriatic arthritis (n=1). 56.1% had stage II, with stage I being the second most common at 29.8%. Elevated CRP was observed in 36.8%, ESR in 33.3% and ACE in 84.2%. 5 patients had positive ANA (8.8%).

Radiological diagnosis was preferably obtained by chest CT (89.5%). Histological diagnosis was reached by lymph node biopsy in 59.6% and by skin biopsy in 33.3%. 54.4% received treatment during some stage of follow-up, preferably with prednisone (49.1%). Azathioprine was associated in 14.6%, methotrexate in 14% and mycophenolate in 8%. 4 patients received biological therapy (3 adalimumab, 1 rituximab). 10.5% suffered some adverse effect related to the treatment that required modification of the regimen. 70.2% of patients are in remission. During follow-up, 7 patients (12.3%) developed neoplasms and 1 (1,7%) died directly from the disease.

Keywords: *sarcoidosis, autoimmune, corticosteroids*

[Abstract:0283]

THE ROLE OF ANTIPHOSPHOLIPID ANTIBODIES IN VENOUS THROMBOEMBOLIC DISEASE: RESULTS OF A PROSPECTIVE STUDY

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Background: Venous thromboembolic disease ("VTE") is the third leading cause of cardiovascular disease and of mortality in the western world. Anti-phospholipid syndrome ("APS") is an acquired pro-thrombotic autoimmune disease that predisposes to thrombotic events and/or obstetric complications, in the persistent presence of anti-phospholipid ("aPL") antibodies. The prevalence of this syndrome in the general population suffering from VTE is around 9%. However, there are few studies that analyze the aPL antibodies prevalence in the acute phase of VTE and their prognosis impact.

Objective: To test classical and extra-criteria aPL antibody positivity in patients suffering from acute VTE.

Methods: A total of 181 patients with acute VTE in the study

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group and 181 persons in the control group were studied, with a two-year follow-up.

Results: The prevalence of classical aPL antibodies was 8.8% and of extra-criteria aPL antibodies was 23.8% in the patient group. Moreover, the development of VTE was significantly associated with IgA aB2GP1 antibody positivity [16% (29) vs. 5% (9); OR 3.6 (95% CI 1.67-7.95); p=0.001]. During the follow-up IgA aB2GP1antibody positivity was associated with the most severe forms of presentation [21% (20) vs. 8% (4), OR 3.13 (95% CI 1.00-9.73); (p=0.048)] and with the development of chronic thromboembolic pulmonary hypertension [50% (5) vs. 14.8% (23); OR 5.7 (95% CI 1.53-21.40); p=0.008].

Conclusions: IgA aB2GP1antibody positivity is a risk factor for suffering VTE, especially severe forms and for the development of chronic thromboembolic pulmonary hypertension in VTE patients.

Keywords: *venous, thromboembolism, antiphospholipid*

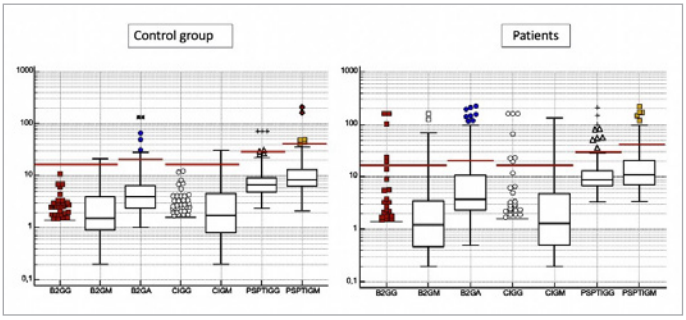


Figure 1. *aPL antibody positivity in VTE.*
Acronyms and abbreviations: B2GG, IgG anti-Beta2 glycoprotein1; B2GM; IgM anti-Beta2 glycoprotein1; B2GA, IgA anti-Beta2 glycoprotein1; CIGG, IgG anti-cardiolipin; CIGM, IgM anti-cardiolipin; PSPTIGG, IgG anti phosphatidylserine prothrombin; PSPTIGM, IgM IgG anti phosphatidylserine prothrombin.

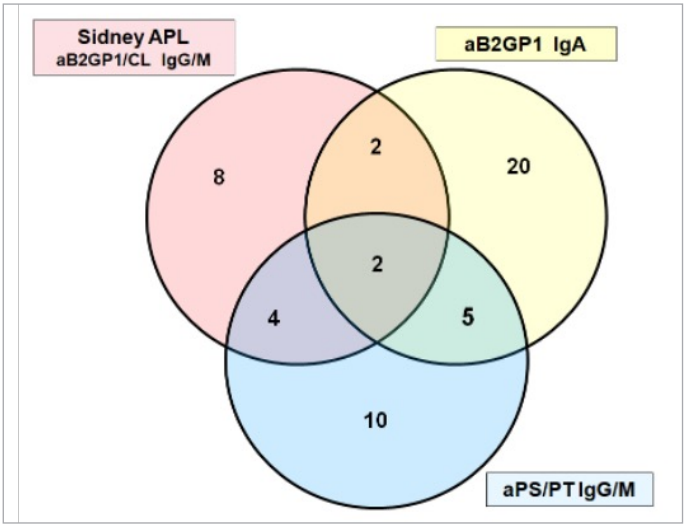


Figure 2. *aPL positivity in VTE.*
Acronyms and abbreviations: aB2GP1 IgA, IgA anti-Beta2 glycoprotein1; aPS/PT IgG/M, IgG and IgM anti phosphatidylserine prothrombin; aB2GP1/CL IGG/M, IgG and IgM anti-Beta2 glycoprotein1 and anti-cardiolipin.

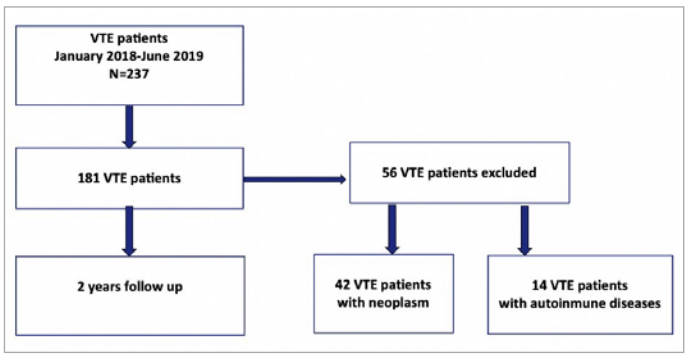


Figure 3. *Disposition algorithm and flow diagram.*
The study group consisted of 181 adult patients with acute VTE, who required admission to the 12 de Octubre University Hospital and were recruited consecutively between January 2018 and June 2019. Acronyms and abbreviations: VTE, venous thromboembolism.

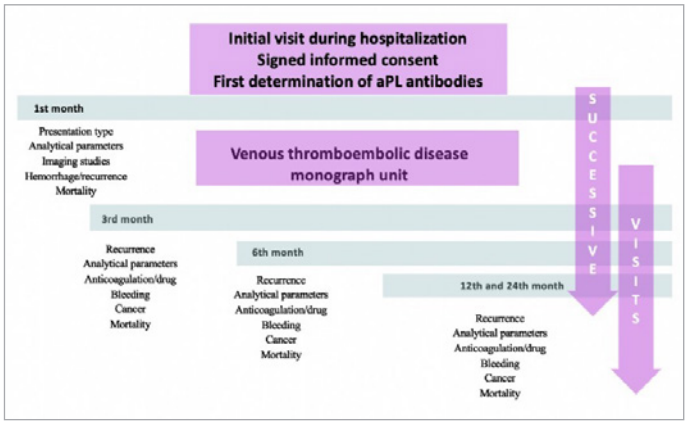


Figure 4. *Variables and evaluation instruments.*
The patients were followed up for a total of 24 months in the thromboembolic disease monographic unit of the Hospital 12 de Octubre.The following variables were collected in the following consultations Form of presentation Analytical parameters Imaging studies Mortality Diagnosis of cancer Hemorrhagic complications Recurrences Thrombotic complications.

INCLUSION CRITERIA
I.1. Age over 18 years old.
I.2. DVT and/or PTE confirmed by ultrasound with venous Doppler and/or angioCT of pulmonary arteries.
I.3. Hospital admission.
I.4. Signature of the informed consent form.
EXCLUSION CRITERIA
E. 1. Suffer an active oncologic disease.
E.2. Suffer a systemic autoimmune disease.

Table 1. *Inclusion and exclusion criteria.*
DVT, deep venous thrombosis; PTE, pulmonary thromboembolism; CT, computed tomography.

aPL antibodies	Control group (n=181)	Patients (n=181)	OR (CI 95%)	p(value)
Classical aPL antibodies	4 (2.2%)	16(8.8%)	4.30(1.40-13.10)	0.010

Table 2. *Classical aPL antibody positivity.*
Classical aPL antibody positivity in the cohort is 8.8% versus 2.2% in the control group.Acronyms and abbreviations: aPL, antiphospholipid.

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aPL antibodies	Control group	Patients	OR(CI 95%)	p(value)
Extra-criteria aPL antibodies	20(11%)	42(23.8%)	2.51(1.41-4.47)	0.001
IgA aB2GP1	9(5%)	29(16%)	3.65(1.67-7.95)	0.001

Table 3. *Extra-criteria aPL antibody positivity.*
Extra-criteria aPL antibody positivity reached 24%. IgA aB2GP1isotype is the only one that show a significant association in patients with VTE over the general population.
Acronyms and abbreviations: IgA aB2GP1, IgA anti-Beta2 glycoprotein1.

Variable	Odds ratio	CI 95%	p(value)
Hypertension	1.22	0.62-2.39	0.560
Obesity	5.54	2.94-10.43	<0.001
Dyslipidemia	1,18	0.61-2.28	0.627
Smoking	2,45	1.21-4.96	0.013
Age over 65	1.51	0.75-3.04	0.257
Classical aPL antibodies	4.61	1.26-16.91	0.021
IgA aB2GP1	3.78	1.47-9.7	0.006
IgM/IgG aPS/PT	0.72	0.26-2.05	0.543
Area under de ROC curve	0.808	0.758-0.851	

Table 4. *aPL positivity and unprovoked VTE. Multivariant analysis.*
Multivariate analysis demonstrates that risk factors independently associated with the development of unprovoked VTE are obesity, and the presence of classical aFL antibodies and of IgA aß2GP1 antibodies.
Acronyms and abbreviations: IgA aB2GP1, IgA anti-Beta2 glycoprotein1, aPS/PT, anti-phosphatidylserine prothrombin.

Variable	CPTEH(n=10)	No CPTEH (n=164)	OR (95% CI)	p(value)
Age(years) Median (IQR)	82 (IQR 79 86)	74(IQR 50 85)	1.06(1-1,12)	0.047
Respiratory insufficiency	7(78%)	43(34%)	6.67(1.32-33.53)	0.021
IgG aB2GP1	0	4(2.6%)	ne	0.999
IgM aB2GP1	0	9(5.8%)	ne	0.999
IgA aB2GP1	5(50%)	30(20%)	5.95(1.59-22.27)	0.008
IgG aCL	0	6(4%)	ne	0.999
IgM aCL	0	8(5.2%)	ne	0.999
IgG aPS/PT	0	7(4.5%)	ne	0.999
IgM aPS/PT	0	16(11%)	ne	0.999
AL	0	12(8.3%)	ne	0.999
Tryple positivity	0	3(2%)	ne	0.999

Table 5. *Chronic thromboembolic pulmonary hypertension.*
During follow-up, 6.1% of patients (10/164) developed CTEPH, with an annual rate of 3.06%. Risk factors significantly associated with the development of CTEPH were: age [median 82 (IQR 79-86) vs. 74 (IQR 50 85) OR 1.06 (95% CI 1-1.12); p=0.04]; the presence of respiratory failure in the acute phase of VTE [77.8% vs. 34.9%; OR 6.52(95% CI 1.29-32.74); P=0.023]; and positivity for IgA aB2GP1 antibodies [50% vs. 14.8%; OR 5.7 (95% CI 1.53-21.40); P=0.008]Acronyms and abbreviations: CPTHE, chronic pulmonary thromboembolism; aB2GP1, anti-Beta2 glycoprotein1, aPS/PT, anti-phosphatidylserine prothrombin; AL, Lupic anticoagulant.

[Abstract:0295]

LUNG-KIDNEY SYNDROME IN THE CONTEXT OF MICROSCOPIC POLYANGIITIS

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Introduction: Vasculitis is a heterogeneous group of diseases resulting in immunological inflammatory reaction with blood vessels walls damage. It can cause organ failure or death; hence timely diagnosis and treatment are necessary.

Clinical Case: a 75-year-old man, with a history of metabolic syndrome, chronic kidney disease, prostate adenocarcinoma, and anemia under study, came to the emergency service due to asthenia and dyspnea with months of evolution and an episode of hemoptysis in small amount after coughing. No other documented hematic losses. Physical examination reported: pale skin, hypertensive, tachycardic and hypoxic. Analytically, severe normochromic normocytic anemia, urea/creatinine 180/4.19mg/dL, iron deficiency, sedimentation rate 130mm, immunoglobulins in normal range, increased haptoglobin, negative Coombs, increased C-reactive protein, and protein/creatinine ratio in the nephrotic range. Chest X-ray with heterogeneous infiltrate in the right lung field and computed tomography showing diffuse alveolar hemorrhage, with superimposed infectious process. Transfusion was performed and antibiotic was started. On D2 of hospitalization, clinical worsening, starting pulses of methylprednisolone due to suspicion of lung-kidney syndrome (LKS) in context of possible vasculitis, transferring the patient to intensive care, where begun plasmapheresis and evolved favorably. Afterwards, the result of remaining study with anti-neutrophil cytoplasmic myeloperoxidase antibody (p-ANCA) positive, assuming a diagnosis of microscopic polyangiitis (MP).

Discussion: MP is a p-ANCA positive small vessel vasculitis, which presents mostly with LKS and can evolve quickly in glomerulonephritis and alveolar hemorrhage. Treatment depends on the severity of the disease and therefore it is essential to act in a timely manner to ensure differentiated treatment for patients.

Keywords: *vasculitis, microscopic polyangiitis, lung-kidney syndrome*

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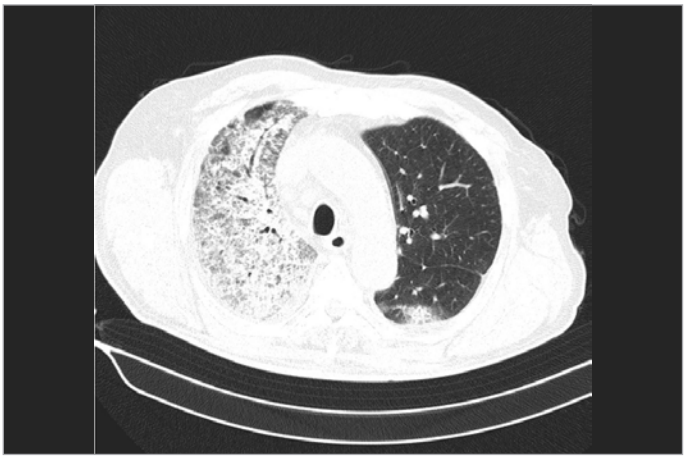


Figure 1. CT scan.



Figure 2. X-ray.

[Abstract:0331]

IL-1B/DNA COMPLEX ELISA AS A SIMPLE DIAGNOSTIC TOOL FOR IL-1-MEDIATED AUTOINFLAMMATORY DISEASES

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Introduction: IL-1-mediated autoinflammatory diseases (AID) are a heterogenous group of innate immunity disorders. Previous studies indicated that the release of neutrophil extracellular traps (NETs) carrying IL-1β on their DNA scaffold is a key pathogenic mechanism. However, the differential diagnosis and timely selection of appropriate therapy remain challenging.

Aim and Methods: This study provides first evidence for a novel "in-house" IL-1β/DNA complex ELISA, as a putative diagnostic method for AID. Measurements were performed in plasma obtained by active, treatment-naïve patients with AID, bacterial infections (INF) and autoimmune/rheumatic diseases (ARD). Sex/age-matched healthy individuals (HI) served as controls. Concomitantly, immunofluorescence microscopy in isolated neutrophils, was used as a gold-standard to detect IL-1β-enriched NETs and verify the results.

Results: In total 25 HI, 38 INF, 19 ARD and 22 AID patients were recruited, according to standard diagnostic criteria. AID group included 12 monogenic disorders (10 FMF, 1 CAPS, 1 PAPA), 8 adult-onset Still's disease and 2 idiopathic recurrent pericarditis. While measurements of free IL-1β in plasma using commercial ELISA, did not differ among groups, AID group yielded significantly higher levels of IL-1β/DNA complexes compared to HI ($p<0.0001$), INF ($p<0.001$), and ARD ($p<0.0001$) groups, that were in consistence with immunofluorescence analysis. Additionally, IL-1β/DNA complex measurements were independent to total cell-free DNA levels (SYTOX-green dye) in plasma. Moreover, in selected AID patients, IL-1β/DNA complex levels significantly reduced after successful response to anti-IL-1 agents.

Conclusions: Assessment of circulating IL-1β/DNA complexes appears to be a novel, simple and fast diagnostic tool for AID. Large-scale, multicenter, validation is needed.

Keywords: *IL-1β, neutrophil extracellular traps, autoinflammatory diseases, ELISA*

[Abstract:0336]

DYSPHAGIA IN INFLAMMATORY MYOPATHIES: CHARACTERISTICS, TREATMENT AND EVOLUTION IN 28 PATIENTS

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Medicina Interna, Hospital Clínico Universitario Virgen de la Victoria, Málaga, Spain

Objective: To evaluate the prevalence, treatment and evolution of dysphagia in inflammatory myopathies.

Materials and Methods: Descriptive and retrospective observational study, in which patients seen in the autoimmune diseases clinic during the last two years, with a probable or definite diagnosis of inflammatory myopathy, have been identified, assessing their pattern of muscle involvement, presence of dysphagia, treatment received and evolution of symptoms.

Results: Out of a total of 28 patients with a diagnosis of

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inflammatory myopathy, 9 had associated dysphagia. Among these we found (Figure 1): 4 dermatomyositis (DM), 2 immune-mediated necrotizing myopathy (IMNM), 1 polymyositis (PM), 1 inclusion body myositis (IBM) and 1 amyopathic dermatomyositis (aDM). 33.3% had dysphagia to liquids, 11.1% to solids and 55.6% to both textures (Figure 2). Among the groups with DM, aDM, PM and IMNM, 100% received treatment with oral prednisone. 25% received pulses of intravenous methylprednisolone and 62.5% received immunoglobulins, both infusions in cases of rapid clinical worsening. Other treatments are included in Figure 3. For the only case of dysphagia associated with IBM, with intrinsic resistance to corticosteroids, we used metrotexate and tacrolimus without total effectiveness.

Conclusions: Dysphagia is a serious problem in the pattern of involvement of inflammatory myopathies, with notable prevalence and a great impact on quality of life. Corticotherapy is the fundamental pillar in its treatment. It would be interesting to investigate new specific therapeutic lines based on the different patterns of involvement.

Keywords: *inflammatory myopathies, dysphagia, autoimmune, corticotherapy*

	Total	DM	PM	IMNM	IBM	aDM
Dysphagia patients (% total)	9 (32%)	4 (14%)	1 (3,5%)	2 (7%)	1 (3,5%)	1 (3,5%)
Sex (Male/Female)	(5/4)	(2/2)	(1/0)	(1/1)	(1/0)	(0/1)
Age, median (interquartile range)	63 (27)	52 (22)	56	82,50 (0)	73	63

Figure 1. *Quantitative variables of the group of patients with inflammatory myopathies.*

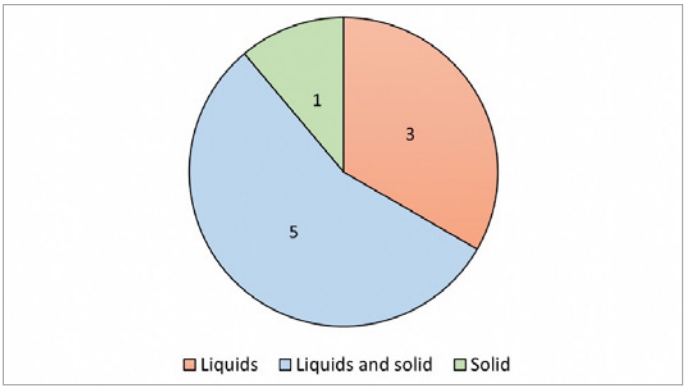


Figure 2. *Distribution of dysphagia type (liquids and solids).*

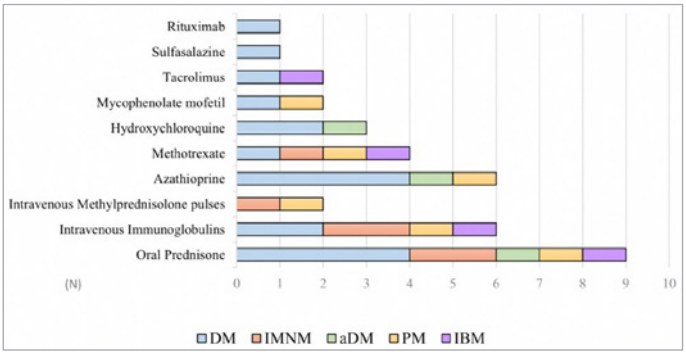


Figure 3. *Treatment lines used for dysphagia in different inflammatory myopathies.*

[Abstract:0338]

FIBROSIS AND EOSINOPHILIA, SUSPICIOUS

Daniel De la Cueva Genovés, Elena Basallote Leal, Jesús Antonio Cívico Ortega, Carla Álvarez González, Eva Romero Polo, Alba Ramírez Bueno, Ana Hidalgo Conde, Rocío Arnedo De Los Ríos

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Endomyocardial fibrosis (EMF) is a rare condition characterized by the formation of fibrous tissue in the myocardium, often linked to various medical conditions. This case presents a 64-year-old woman with a history of familial mediterranean fever under treatment with colchicine. He was admitted with suspicion of acute pericarditis due to pleuritic pain in the left hemithorax of two weeks' duration.

During admission, no electrocardiographic alterations indicative of ischemic heart disease or acute pericarditis were observed, but persistent eosinophilia and troponin elevation were notable. Coronary angiography was normal, and echocardiography revealed thickening in the inferior and inferolateral septal basal segments, indicative of EMF, confirmed by CT scan (Figure 1) and Cardio MRI (Figure 2). The CT scan showed vasculitic lesions in the abdomen (Figure 3) and skull (Figure 4), possibly embolisms related to the myocardial findings.

Discharged with corticosteroids and anticoagulants, she returned with neurological worsening a month later. The study revealed brain lesions that were not clearly embolic and new granulomatous skin lesions. Relating the findings of EMF, eosinophilia and granulomatous dermatitis, the established diagnosis was Eosinophilic Granulomatosis with Polyangiitis (EGPA). Treated with corticosteroids and immunosuppressants, she experienced great clinical improvement.

This case highlights the association of EMF with EGPA, underscoring the need to rule out this diagnosis after presentation. Coexistence raises questions about the relationship between eosinophilic inflammation and myocardial fibrosis, complicating treatment, although similar to that of EGPA, with corticosteroids and immunosuppressants. The patient, after months of treatment, had resolution of the skin lesions and regression of endomyocardial fibrosis.

Keywords: *endomyocardial fibrosis, EMF, eosinophilic granulomatosis with polyangiitis, EGPA, granulomatous dermatitis*

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Figure 1. *(Heart CT) Myocardial thickening in the inferior septal and inferolateral basal segments.*

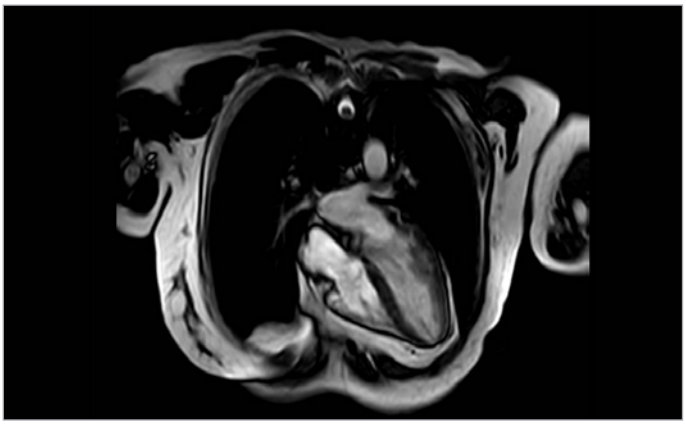


Figure 2. *(Heart MRI) Myocardial thickening in the inferior septal and inferolateral basal segments.*

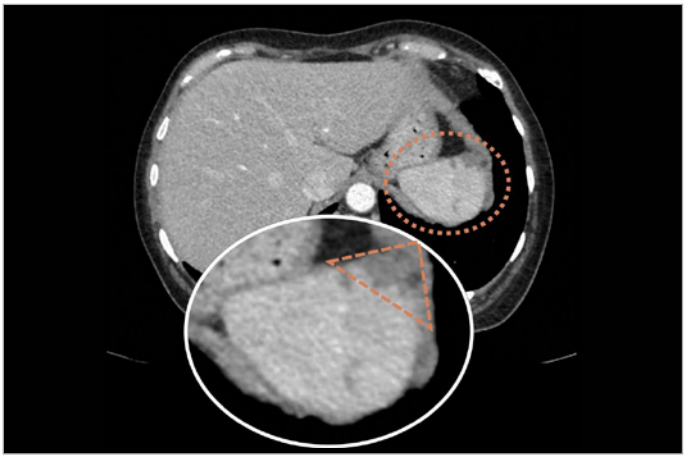


Figure 3. *(Abdominal and Pelvic CT) Small peripheral hypodense lesions with a triangular appearance, suggestive of small splenic infarcts.*

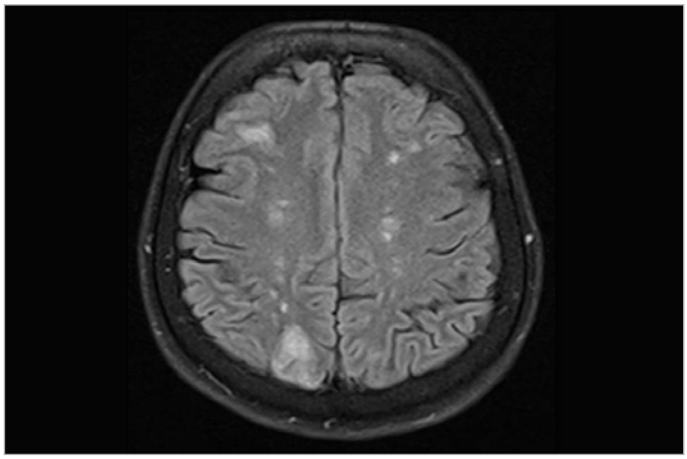


Figure 4. *(Head CT) Acute bihemispheric ischemic lesions.*

[Abstract:0343]

PATIENT WITH HYPERPIGMENTATION, ULCERATED LESIONS AND WEIGHT LOSS

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A 49-year-old female patient presented with complaints of skin darkening, erythematous areas of the skin, ulcerated lesions, and excoriations due to itching, difficulty swallowing, widespread body pain, and 17 kg weight loss in the last 6 months. Muscle strength was found to be 3-4/5 in all extremities. Normocytic anemia, eosinophilia, elevated lactate dehydrogenase, and erythrocyte sedimentation rate were observed. Creatinine kinase and C-reactive protein were within the normal range. Rheumatologic markers were ordered. ANA 1/320, speckled pattern positive. A skin biopsy was done and melanophages were observed. Because of severe tiredness and hyperpigmentation; a basal cortisol test was done. Basal cortisol level was sufficient to rule out adrenal insufficiency. Because of constitutional symptoms and elevated LDH, solid and hematologic malignancies were screened. Atypical cells were seen in the peripheral blood smear and a bone marrow biopsy was performed. Malignancy and infiltration were not detected in the bone marrow biopsy. An excisional biopsy of the axillary lymph node was done and dermatopathic lymphadenopathy was observed. No evidence of malignancy was found in CTs of neck-thorax-abdomen, mammography, gynecologic examination, and endoscopic examinations. Because of muscle weakness, an MRI and EMG were ordered. Signal increases in STIR sequence and mild contrast enhancements in muscles on thigh magnetic resonance imaging. EMG revealed diffuse myogenous involvement. Deltoid muscle biopsy revealed atrophic fibers, necrosis, and mononuclear inflammatory infiltration. Dermatomyositis was diagnosed. A myositis-specific antibody panel was sent and Anti-SAE1 was positive. Methylprednisolone, methotrexate, and hydroxychloroquine were started; symptoms and signs of anti-SAE1 dermatomyositis were improved.

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Keywords: *dermatomyositis, myositis-spesicfic antibody, anti-SAE1,*



Figure 1. *Hyperpigmentation on the face.*



Figure 2. *Ulcerated lesions on the right hand.*

[Abstract:0346]

CANCER IMMUNOTHERAPY ADVERSE EVENTS AS A MISSING LINK BETWEEN POLYARTHRITIS AND WORSENING DIABETES: CASE REPORT

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Case Description: A 63-year-old man was admitted in emergency room with decompensated diabetes, swollen left knee and polyarthralgia. Few days earlier, he reported joint symptoms and worsening of glycaemic control that required switch to insulin from metformin. His past medical history was significant for relapsing bladder cancer, for which he had been taking pembrolizumab, an immune checkpoint inhibitor (ICI), for 2 months. At admission, joint examination detected tender left knee, left shoulder functional limitation, bilateral positive squeeze test. Laboratory studies highlighted: WBC 7500/μL, CRP 7.3 mg/dL, glycaemia 350 mg/dL, C-peptide <0.1 ng/mL, HbA1c 8.5%, no ketonuria.

Clinical Hypothesis: Immune-related adverse events triggered by ICI.

Diagnostic Pathways: Musculoskeletal ultrasound revealed joint effusion and synovial hypertrophy of left knee, bursitis of left shoulder, minimal effusion without synovitis of some joints of the hands; rheumatoid factor and anti-CCP resulted negative. Knee arthrocentesis showed inflammatory fluid; neither crystals nor bacteria were detected. Asymmetric involvement and negative serology made the diagnosis of rheumatoid arthritis unlikely; synovial fluid analysis was not consistent with septic arthritis or crystal arthropathy. Onset of symptoms after immunotherapy suggested diagnosis of ICI-induced arthritis; at the same time, low levels of C-peptide and poor glycaemic control supported the hypothesis of ICI-induced diabetes mellitus.

Discussion and Learning Points: ICI-induced arthritis usually presents with seronegative polyarthritis. It is treated with NSAIDs, corticosteroids or immunosuppressive drugs, depending on severity. ICI-induced diabetes mellitus is characterized by presentation in diabetic ketoacidosis or extremely low C-peptide. It can occur despite pre-existing diabetes and requires insulin therapy.

Keywords: *immune checkpoint inhibitors, immune related adverse events, ICI-induced arthritis, ICI-induced diabetes mellitus*

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[Abstract:0348]

IL-1B-ENRICHED NEUTROPHIL EXTRACELLULAR TRAPS AS A MARKER OF INFLAMMATION AND RENAL TISSUE DAMAGE IN FAMILIAL MEDITERRANEAN FEVER-ASSOCIATED AMYLOIDOSIS: A CASE STUDY

Christina Antoniadou[1], *Anastasia Maria Natsi*[2], *Efstratios Gavriilidis*[1], *Victoria Tsironidou*[2], *Pelagia Kriki*[3], *Georgios Liapis*[4], *Konstantinos Ritis*[1], *Panagiotis Skendros*[1]

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Background and Aims: IL-1β plays a pivotal role in inflammation and is a target of treatment in familial Mediterranean fever (FMF). Previously, we demonstrated that peripheral neutrophils release IL-1β-bearing neutrophil extracellular traps (NETs) during disease attacks. In this case, we provide the first evidence that the neutrophils/NETs/IL-1β axis may be involved in FMF-associated amyloidosis.

Methods: A 52-year-old man was referred to our hospital with severe nephrotic syndrome. He reported two decades of self-limited recurrent attacks involving fever, abdominal pain, and pleurisy. MEFV gene mutation analysis and kidney biopsy were performed. Additionally, markers of NET formation and IL-1β production were assessed in peripheral neutrophils, plasma, and renal tissue.

Results: The detection of the heterozygous *M694V* mutation confirmed the clinical diagnosis of FMF. Kidney histopathology and electron microscopy imaging were compatible with AA amyloidosis. Elevated levels of circulating IL-1β/DNA complexes were consistent with immunofluorescence microscopy analysis, indicating the simultaneous presence of IL-1β-enriched NETs in peripheral neutrophils and renal tissue. Due to the partial response to colchicine and the persistence of increased plasma levels of IL-1β/DNA complexes, anti-IL-1β treatment with canakinumab was commenced. A favorable response to canakinumab was accompanied by a significant, concomitant reduction of proteinuria and IL-1β+ NETs markers in peripheral neutrophils and plasma, which has been maintained for 18 months post-treatment.

Conclusions: In FMF, IL-1β-enriched NETs may contribute to chronic inflammation leading to organ tissue damage, such as amyloidosis. Markers of the neutrophils/NETs/IL-1β axis could be useful for the differential diagnosis and timely treatment selection in autoinflammatory disorders.

Keywords: *familial mediterranean fever, amyloidosis, neutrophil extracellular traps, IL-1β, canakinumab, autoinflammatory diseases*

[Abstract:0359]

BEYOND HIVES… WHEN MONOCLONAL GAMMOPATHY HAS A CERTAIN MEANING

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Schnitzler's syndrome is a rare systemic disease, likely acquired, that could be classified among autoinflammatory syndromes due to shared characteristics. This syndrome is characterized by recurrent febrile rash, arthromyalgias, adenopathies, leukocytosis, and a systemic inflammatory response. The defining features of the disease include monoclonal gammopathy of the IgM type and chronic urticarial rash. The prevalence of the disease is unknown, with an estimated 150 reported cases, mostly in Europe (four of them at our hospital.

Among the four patients at the Hospital Fundación Jiménez Díaz (Madrid), all of them were women with the following ages: 91 years (case 1), 43 years (case 2), 55 years (case 3), and 51 years (case 4). The onset of symptoms occurred at 37, 19, 54, and the first year of life, respectively. Three of them were diagnosed in 2023, while case 1 was diagnosed in 2015. All of them exhibited urticarial rash and elevated IgM, except for case 3, who presented an IgA monoclonal component. Articular involvement was observed in 75% of the cases, fever in 50%, leukocytosis in 50%, C-reactive protein and erythrocyte sedimentation rate were elevated in 25%. Anakinra (anti-IL-1) was the initial treatment (case 4 has not started yet), showing a positive response in all of them, except for case 3, which casts doubt on the primary diagnosis.

Therefore, we present a description of four cases of Schnitzler's syndrome, highlighting their similarities and differences. This presentation may contribute to the diagnosis of the disease and offer insights into future therapeutic options.

Keywords: *Schnitzler syndrome, anti IL-1, autoinflamatory syndrome, IgM*

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[Abstract:0396]

A CASE OF GOODPASTURE'S SYNDROME, CYTOMEGALOVIRUS AND ALVEOLAR HAEMORRHAGES: WHEN VASCULITIS IS NOT EVERYTHING

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The anti-glomerular basement membrane disease (formerly Goodpasture Syndrome) affects 0.5-1 case/million- year, mainly in the 3th-7th decades. It is a small-vessel vasculitis caused by anti-MBG autoantibodies depositing in the glomerular and/or alveolar basement membrane, triggering a neutrophil-dependent inflammatory reaction, leading to the disease's main characteristics: rapidly-progressive glomerulonephritis and alveolar hemorrhage.

An 18-year-old male, with no significant medical history, presented to the Emergency Department with a 24-hour history of asthenia and hemoptysis, exhibiting mucocutaneous pallor and generalized hypophonesis on auscultation. Blood tests revealed 6 mg/dl of hemoglobin and 3.51 mg/dl of creatinine, along with bilateral alveolar infiltrates on chest X-ray; CT-scan confirmed alveolar hemorrhage.

Antibody studies were conducted, only anti-MBG was positive. Renal biopsy was delayed, prioritizing high-dose corticosteroids, cyclophosphamide, and plasma exchange, leading to ICU admission.

During intensive care stay, the patient's respiratory status worsened, and Cytomegalovirus was isolated from blood and bronchoalveolar lavage, sequently treated with Ganciclovir. Despite an initial improvement, cytopenias due to cyclophosphamide appeared, leading to a switch to foscarnet. After progressive worsening and recurrence of alveolar hemorrhage, treatment with rituximab was considered. After 41 days, the patient died.

Cytomegalovirus is a common infection in glomerulonephritis patients treated with cyclophosphamide1 but there are only a few cases of relapsing alveolar hemorrhage in anti-GBM due to cytomegalovirus 2,3. The fact that the patient did not improve with immunosupression supports this notion, and the optimal treatment with Ganciclovir was held because of the cytopenias. Therefore, given the severity of the disease, a high suspicion and prompt diagnostic are crucial.

Keywords: *Goodpasture syndrome, alveolar hemorrhages, cytomegalovirus, anti-MBG*

[Abstract:0400]

RECURRENT ANTERIOR UVEITIS

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Case Description: A 32-year-old male, native of Paraguay, resident in Spain for 6 years, who was referred to the Internal Medicine Outpatient Clinic from ophthalmology for 3 episodes of bilateral anterior uveitis in the last 7 months. In the anamnesis, the patient denied systemic symptoms. Physical examination showed splenomegaly. Laboratory tests showed a pattern of dissociated cholestasis, and the rest of the serology, autoimmunity, complement, immunoglobulins, proteinogram, RCT and Mantoux were normal. Abdominal ultrasound showed splenomegaly and a thoracic CT scan showed bilateral axillary, prevascular and paratracheal right upper and lower right and lower right and left hilar subcentimetric lymphadenopathies, as well as a pulmonary micronodular pattern.

Clinical Hypothesis: We were therefore faced with a patient with bilateral chronic anterior anterior uveitis, adenomegaly, splenomegaly and pulmonary interstitial pattern, in whose main differential diagnosis we raised the possibilities of sarcoidosis, tuberculosis and neoplasia.

Diagnostic Pathways: Finally, it was decided to perform a liver biopsy with histological diagnosis of non-caseating epithelioid granulomas, so he was diagnosed with systemic sarcoidosis.

Discussion and Learning Points: Sarcoidosis is a granulomatous disease that mainly affects young adults, with female predominance. The clinical manifestations are very diverse, as any organ can be affected. The natural history is highly variable, with two thirds of patients presenting with spontaneous remission and 10-30% with a chronic course. The diagnosis is established by the existence of non-calcifying epithelioid granulomas in one or more organs or systems, with other causes of granulomatous disease previously excluded.

Keywords: *uveitis, splenomegaly, lymphadenopathy, sarcoidosis.*

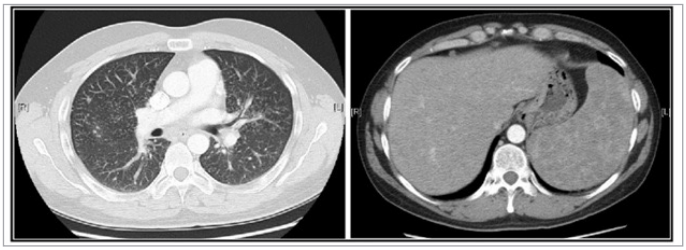


Figure 1. *Micronodular pattern, lymphadenopathy and splenomegaly. Thoracic CT scan showed bilateral axillary, prevascular and paratracheal right upper and lower axillary and right and left hilar subcentimetric lymphadenopathy, as well as pulmonary micronodular pattern and splenomegaly.*

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[Abstract:0406]

THE TREATMENT ROAD FOR GASTRIC ANTRAL VASCULAR ECTASIA IN THE SETTING OF SYSTEMIC SCLEROSIS

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Gastric antral vascular ectasia (GAVE) is a rare condition occurring in systemic sclerosis (SSc) patients, characterized by dilated stomach blood vessels leading to chronic blood loss.

A 62-year-old woman with diffuse SSc and interstitial lung disease (ILD) presented severe anemia and GAVE, initially marked by a hemoglobin level of 4.9 g/dL and a modified Rodnan skin score (mRSS) of 24. The initial endoscopic examination revealed prominent erythematous stripes known as "watermelon stomach" (Figure 1A), with positive ANA (1:160, nuclear speckled), anti-polymerase III (+), and anti-Ro/SSA (+).

Within three months, the patient experienced rapidly progressing skin thickening and tendon friction rubs in the fingers, wrists, and elbows (Figure 2). Argon plasma coagulation (APC) was applied thrice weekly for 15 sessions to address bleeding, in conjunction with Tocilizumab for the treatment of SSc with ILD. Tocilizumab was discontinued due to side effects, including neutropenia and thrombocytopenia. This led to the initiation of intravenous immunoglobulin (IVIG) treatment for four months (at a dosage of 1g/kg per month), followed by mycophenolate mofetil at 2000mg/day (Figure 3). The combination of immune-modulating agents with APC demonstrated efficacy, particularly in stabilizing hemoglobin levels when bleeding persisted after endoscopic coagulation.

After five months, the patient's anemia and GAVE improved, reflected in a hemoglobin level of 9.9 g/dL and resolved GAVE lesions on follow-up endoscopy (Figure 1B-D). This case emphasizes the successful management of GAVE through repeated APC sessions and concurrent use of immune-modulating agents in a patient with early progressive SSc.

Keywords: *systemic sclerosis (SSc), gastric antral vascular ectasia (GAVE), argon plasma coagulation (APC), tocilizumab, Intravenous immunoglobulin (IVIG)*

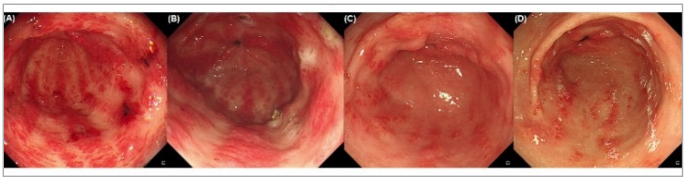


Figure 1. *Endoscopic picutures of the gastric antrum of the patient over time. (A) Initial (B) A week later (C) 3 months later (D) 5 months later.*



Figure 2. *Sclerodactyly of both hands of the patient.*

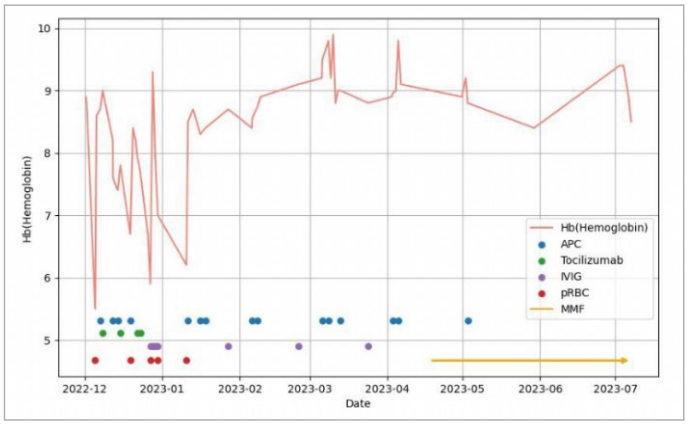


Figure 3. *Overall course of treatment.*

[Abstract:0411]

A CASE OF AUTOIMMUNE HEPATITIS

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Introduction: Autoimmune hepatitis (AIH) is a chronic inflammatory liver disease characterized by circulating autoantibodies and elevated serum globulin levels, with a variable clinical spectrum.

Case: Female patient, 44 years old, without pathological history, went to the emergency department due to asthenia, choluria and jaundice that had lasted 1 week. She denied consuming alcohol, drugs, or medication. Analytically there was an increase in transaminases with marked hyperbilirubinemia, due to direct bilirubin. The abdominal CT scan showed slight homogeneous splenomegaly. The patient was hospitalized with of acute hepatitis of unclear etiology. The study showed: negativity for hepatotrophic virus serology, marked complement consumption, positive AMA M2 and positive LC1. The liver biopsy showed findings compatible with autoimmune hepatitis. In the following two months, due to the persistence of complaints, with concomitant worsening of cytolysis, she was readmitted in the context of AIH with probable overlap with primary biliary cirrhosis (PBC). She repeated the

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biopsy, which showed a liver with architecture that already had some distortion due to the presence of significant fibrosis, predicting a likely cirrhogenic evolution. Upon readmission, she presented a favorable clinical evolution. She was medicated with prednisolone 40 mg/id, ursodeoxycholic acid 500 mg/bid, azathioprim 50 mg/bid and respective prophylaxis.

Discussion: Autoimmune hepatitis can occur at any age and is more frequently in females. The incidence in Europe is estimated to be 0.9 to 2 cases per 100,000 inhabitants per year. Given the variable clinical spectrum, it's important to exclude other etiologies as well as consider overlap with other autoimmune diseases.

Keywords: *autimmune hepatitis, primary biliary cirrhosis, cirrhosis, transaminases*

[Abstract:0439]

DECIPHERING THE PUZZLE: DOPPLER ULTRASOUND AS THE KEY TO DIAGNOSING TEMPORAL ARTERITIS IN A 73-YEAR-OLD PATIENT IN A RESOURCE-LIMITED HOSPITAL IN CAMEROON

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Introduction: A 73-year-old male presented at a hospital in Cameroon, displaying symptoms indicative of a middle cerebral artery stroke: sensory aphasia, right hemiparesis, and right hemianopsia. His wife reported intense temporal headaches, jaw claudication, and vision loss over two months. Faced with this clinical scenario, a practical diagnostic approach was pursued within the constraints of the Cameroonian healthcare system.

Methods: Within Cameroon's private healthcare system, where patients cover all medical expenses, Notre Dame de la Santé hospital, though resource-limited, offered basic services including a laboratory, microbiology, chest X-ray, and ultrasound, lacking advanced tests like CT scans. To address the need for cost-effective diagnostic options, Doppler ultrasound was chosen.

Results: Temporal arteritis suspicion was confirmed via Doppler ultrasound of the temporal arteries, revealing a hypoechoic halo of 0.8mm on the left and 0.76mm on the right. Concordance between ultrasound findings, clinical symptoms, and an elevated erythrocyte sedimentation rate of 120mm/h supported the diagnosis. Following high-dose methylprednisolone pulses, the patient experienced significant improvement within 24 hours, with resolution of motor deficits, language recovery, and headache cessation.

Discussion: Doppler ultrasound proved valuable in this case. Previous studies, like Schafer et al., substantiated its efficacy by establishing cut-off points for intima-media thickness in temporal arteries (0.65mm; 100% sensitivity and specificity). In a 2023 meta-analysis by Nakajima et al., pooled sensitivity for halo sign was 84%, and specificity was 93%.

Conclusions: In summary, Doppler ultrasound emerges as an economical, accessible and accurate diagnostic method for temporal arteritis, especially in healthcare centers in resource-limited countries.

Keywords: *Doppler ultrasound, temporal arteritis, halo sign*

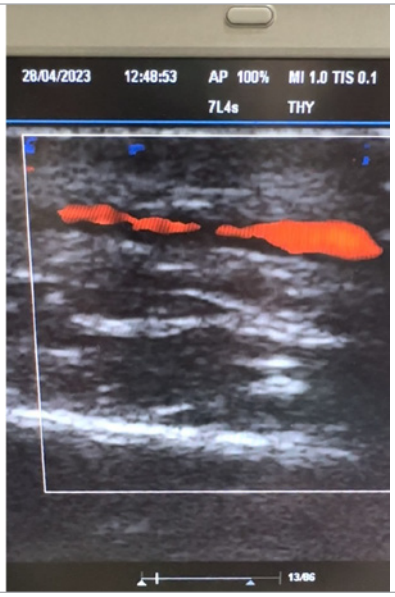


Figure 1. *Halo sign in the left superficial temporal artery in a patient with clinical symptoms consistent with temporal arteritis.*

[Abstract:0447]

ANTI-NUCLEOLAR ORGANIZER REGION 90 ANTIBODIES (NOR90) IN SYSTEMIC SCLEROSIS

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Background: Antinuclear antibodies (ANA) are arguably the most important test of the clinical immunology workup, and the interest for the rare ANA significance is continuously increasing.

Purpose: To analyze the clinical associations of Anti-Nucleolar Organizer Region 90 antibodies (NOR90), rare ANA, in Systemic Sclerosis (SSc).

Methods: We performed a cross-sectional study with retrospective analysis of prospectively collected data by the European Scleroderma Trials and Research (EUSTAR) participating centers, with addition information on NOR90, tested by immunoassays.

Further, we performed a systematic literature search, using the terms "systemic sclerosis" and "NOR90" across three databases: Medline, Scopus, and Web of Science, from inception to November 1st 2023 (Figure 1).

Findings: Overall, data of 1318 SSc patients tested for NOR90 were analyzed from our dataset, 81.3% female with mean age 58.3±13.7 years, of whom 44 (3.3%) were positive for NOR90.

NOR90 were associated with less extensive skin involvement (assessed by the modified Rodnan skin score) and less upper and lower gastrointestinal symptoms, OR(95%CI): 0.376 (0.167-0.845), p=0.018, 0.389 (0.198-0.762), p=0.006 and 0.173 (0.060-0.496), p=0.001, respectively (Figure 2).

No other significant association of NOR90 with SSc-related organ or system involvement was identified.

The literature search identified 17 articles, including a total number of 87 NOR90-positive out of 3357 SSc patients tested, corresponding to an overall prevalence of 2.6%.

Conclusions: To our best knowledge, we present data from the largest SSc cohort tested for NOR90 so far. Our research adds to the previously published studies important data regarding the clinical correlates of these rare ANA.

Keywords: *NOR90, antinuclear antibodies, systemic sclerosis, rare antibodies*

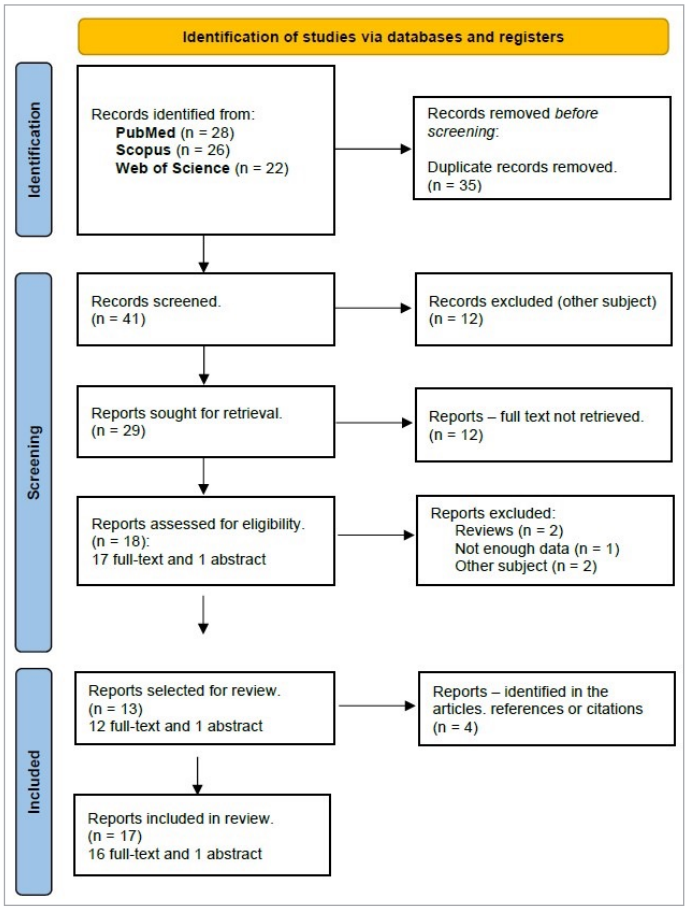


Figure 1. *PRISMA flow chart for literature systematic search: "Systemic sclerosis" AND "NOR90".*

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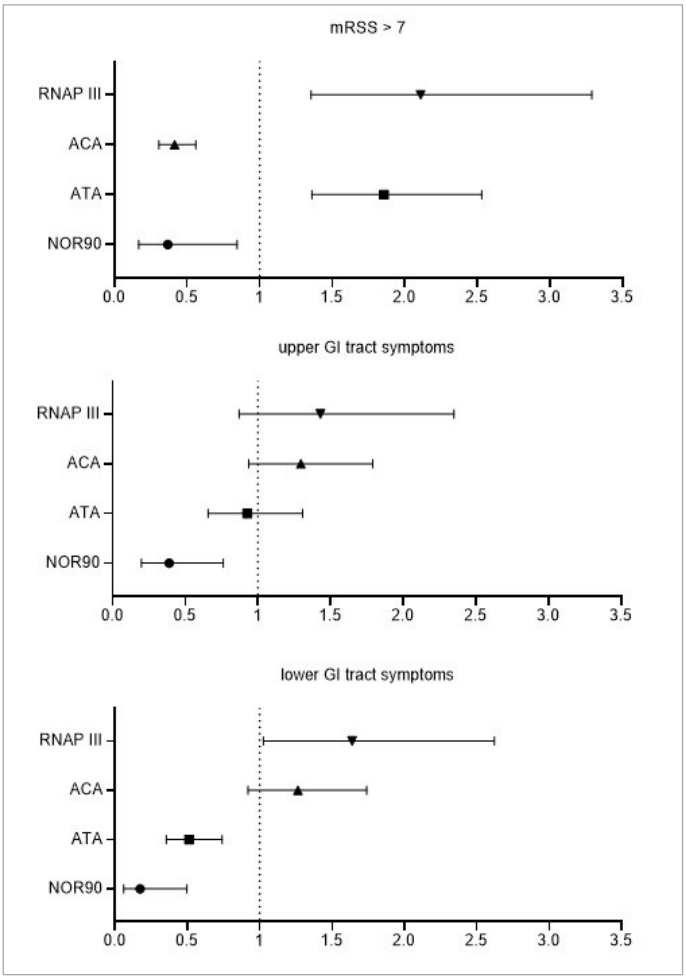


Figure 2. Association of NOR90 and SSc-criteria abs (ATA; ACA; RNAP III) with the mRSS and GI symptoms.

[Abstract:0463]

CHRONIC DIARRHEA IN COMMON VARIABLE IMMUNODEFICIENCY: A DIAGNOSTIC AND THERAPEUTIC CHALLENGE

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Clinical Case: 51-year-old man, with no relevant personal past history, presented with persistent chronic diarrhea, abdominal pain and weight loss in the last year.

Clinical Hypothesis: Infectious and endocrine causes were excluded. Viral serologies and autoimmunity panel were negative. Common Variable Immunodeficiency (CVID) was identified (IgG 451 mg/dL, IgM<25 mg/dL, IgA<6 mg/dL), associated with cobalamin deficiency anemia. Endoscopic exams documented intestinal lymphoid nodular hyperplasia.

Diagnostic Pathways: He was consulted by Autoimmune Diseases and started on endovenous Immunoglobulin without clinical changes. Later, he completed a course of prednisolone with improvement of gastrointestinal symptoms.

Discussion: CVID is a primary immunodeficiency associated with recurrent infections, neoplasms and autoimmune diseases. Chronic enteropathy has been identified in 10 to 20% of patients with CVID, potentiating life-threatening conditions, like malabsorption syndromes and weight loss. Imunoglobulin replacement has shown no efficacy in controlling gastrointestinal symptoms, improvement has only been reported with corticotherapy/immunosuppressors. We describe a CVID-induced enteropathy that improved under corticotherapy. Further extensive studies are needed to prove the effectiveness and more appropriate treatment for this condition, since literature is limited to case reports. With this case, we also intend to represent gastrointestinal involvement in CVID, highlighting the importance of including immunodeficiency as a differential diagnosis of chronic and non-infectious diarrhea, even in the absence of repeated infections.

Keywords: CVID, immunodeficiency, diarrhea

[Abstract:0468]

ANTISYNTHETASE SYNDROME. EARLY MANAGEMENT OF PULMONARY INVOLVEMENT, A CASE REPORT

Elena Urízar

Elena Urízar Ursúa, Alex García Tellado, María Bayona Sánchez, Alicia Aldea Abad, Patricia Marín Oliván, Raúl Parra Fariñas, Joy Selenea Osorio Chavez, Joaquina Lopez-Casas Giner

This clinical report details a 55-year-old male with antisynthetase syndrome (ASS), an uncommon condition with cutaneous and pulmonary manifestations. The patient had a history of hypertension, type 2 diabetes, and type I MEN syndrome with a pituitary macroadenoma. He also suffered recurrent pancreatitis due to hypertriglyceridemia.

In the last two week, the patient developed dyspnea, non-productive cough and skin lesions. He also related bilateral weakness in the pelvic girdle without any other systemic symptoms. Examination revealed afebrile status, mild tachypnea and oxygen dependence. Erythematous plaques, telangiectasias, and hyperkeratosis were observed. Pulmonary auscultation indicated crackles up to the middle lung fields. Laboratory results showed elevated LDH, CK, aldolase, and ESR with negative tumor markers. Chest CT confirmed organizing pneumonia, and PET-CT revealed no significant findings. Autoimmune studies showed positive anti-Ro52 antibodies, supporting the diagnosis of ASS based on cutaneous (mechanic's hands, inflammatory dermatomyositis) and pulmonary involvement (organizing pneumonia).

Intravenous corticosteroid therapy was initiated with a positive clinical response. ASS diagnosis, primarily clinical, poses challenges due to asynchronous manifestations. Pulmonary involvement has a high mortality rate, emphasizing the importance of excluding

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infections before corticosteroid and immunosuppressive treatment. Tumor pathology must be ruled out to prevent misdiagnosis as a paraneoplastic syndrome. Despite negative antisynthetase antibodies, the presence of anti-Ro52, seen in 37% of dermatomyositis cases, indicated a poor response to immunosuppression (1).

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Keywords: *antisyntheatase syndrome, dyspnea, anti-Ro52*

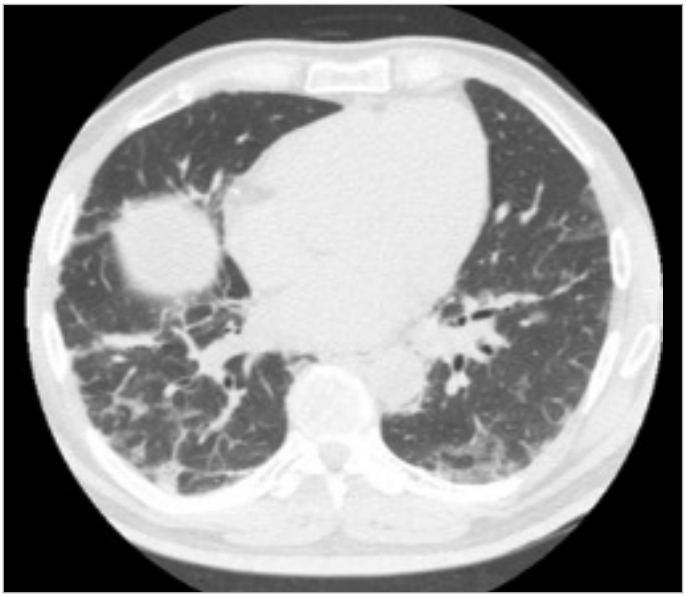


Figure 1. *Pulmonary injury.*

[Abstract:0470]

NEUROSARCOIDOSIS: A GREAT DIAGNOSTIC CHALLENGE. A CASE REPORT

Elena Urízar Ursúa, Ana Serrano Combarro, Carmen Lasa Teja, María Bayona Sánchez, Alicia Aldea Abad, Patricia Marín Oliván

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A 28-year-old male with a history of ADHD and regular marijuana use presented with a first seizure episode. He had recent augmentin treatment for a sudden right retroauricular tumor, diagnosed as a reactive lymph node. Afebrile on examination, the only neurological impairment was bradypsychia. No skin lesions were observed, except for the mentioned lump. Laboratory results indicated elevated CK and acute-phase reactants, and brain imaging revealed enhancements in both temporal lobes. Whole-body imaging showed bilateral mediastinal adenopathies. Infection was ruled out through various tests, and cerebrospinal fluid analysis was normal. A biopsy of the retroauricular lymph node confirmed neurosarcoidosis, prompting corticosteroid and methotrexate treatment. Neurosarcoidosis central nervous system involvement is rare and diagnostically challenging, often resembling other conditions. Cerebrospinal fluid analysis is vital for diagnosis, excluding infectious and tumoral causes. While cranial neuropathy, particularly facial paralysis, is common, sarcoidosis can manifest diversely, as illustrated by seizures in this case. While histological confirmation from a CNS biopsy ensures diagnosis, extracranial biopsy may be acceptable due to technical difficulties. In cases resistant to corticosteroids, early introduction of other immunosuppressants is crucial for disease control.

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Keywords: *neurosarcoidosis, seizure, adenopathies*

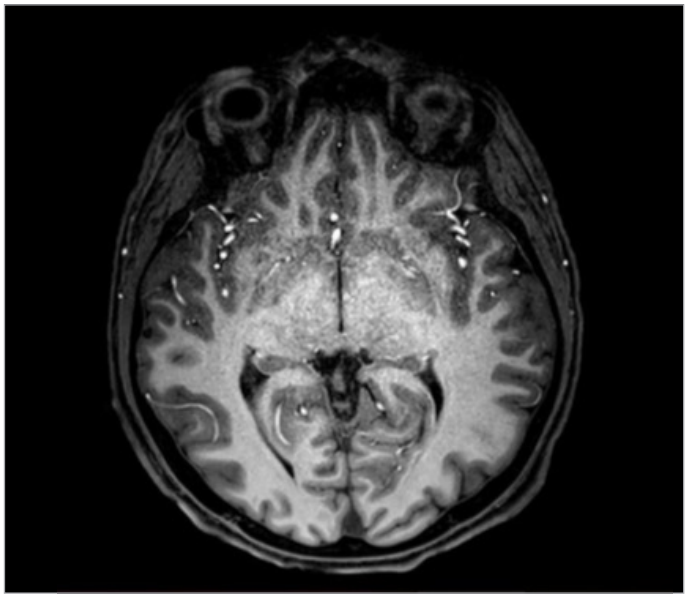


Figure 1. *Brain injury.*

[Abstract:0472]

EOSINOPHILIC FASCIITIS: ABOUT SIX CASES

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Eosinophilic Fasciitis (EF) is a rare condition with uncertain causes and varied clinical presentations. This study aimed to retrospectively examine EF cases in a tertiary-level university hospital's Internal Medicine Department. Demographic, clinical, laboratory data, and treatments were collected from electronic

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medical records. Among 83% female patients, with an average age of 55.3 years at diagnosis, none had hematological or solid organ neoplasms, systemic autoimmune disease, chronic infection, were on hemodialysis, or had intense physical activity before diagnosis. The mean delay in diagnosis was 7 months. Patients primarily exhibited skin-related issues, such as edema and morphea-like lesions on extremities and trunk. Few displayed Groove sign, arthritis, or cubital neuropathy. Notably, there were no myalgias, weakness, or organic complications reported. 50% had eosinophilia, with minimal elevation in acute-phase reactants, and a single patient showed ANA detection at 1/320. Imaging (MRI or ultrasound) displayed widespread thickening of superficial and deep fascia in 83% of cases. Histopathological findings from deep biopsies commonly revealed subcutaneous cellular tissue fibrosis and fascial thickening due to inflammatory infiltration. Treatment predominantly involved varied corticosteroid doses, with some patients needing immunosuppressants or biological therapies due to inadequate responses. Over an average 4.5-year follow-up, all patients showed improvements in cutaneous symptoms without treatment-related complications. In conclusion, despite EF's clinical diversity, all patients in this study predominantly showcased skin-related symptoms resembling edema or morphea. No evident associations with other diseases were found. Diagnosis often involved imaging and deep biopsies, with corticosteroid therapy as the initial choice, complemented by other therapies as needed.

Keywords: *eosinophilic fasciitis, autoimmune diseases, morphea, Eosinophilia*

[Abstract:0490]

MULTIPLE AUTOIMMUNE SYNDROME MAS: REPORT OF A CASE

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Multiple Autoimmune Syndrome (MAS) is a rare and unique entity defined by the presence of at least three autoimmune pathologies in the same individual. There are three types of MAS depending on the associated diseases.

We report the case of a 62-year-old woman with vitiligo and ulcerative colitis for two years treated with mesalazine. Her son also has vitiligo and is being treated for Biermer's disease. During the course of her illness,the patient developed Systemic Lupus Erythematosus, manifesting hematological damage including bicytopenia progressing to agranulocytopenia, normocytic anemia, photosensibility and psoriasiform skin lesions in photoexposed areas, other symptoms included arthralgia, positive ANA at 1/320, and positivity for anti-SSA and anti-SSB antibodies. Associated Sjogren's syndrome presented with severe dry eyes and lymphocytic sialadenitis classified as stage 4 according to Chisholm and Masson. Hashimoto's thyroiditis was also identified with hypothyroidism, positive anti-TPO at 800 IU/ml, goiter, and thyroiditis observed on ultrasound. Stigmata of Biermer's disease included B12 hypovitaminosis with Hunter's glossitis, paresthesias and fundic atrophic gastritis.

The patient received corticosteroids at 1mg/kg along with adjuvant therapy and a gradual tapering, hydroxychloroquine, external photoprotection, thyroid hormone replacement therapy, and intramuscular vitamin B12 per week, followed by monthly administration. The treatment led to normalized blood parameters improvement in overall skin and neurological symptoms.

Multiple Autoimmune Syndrome (MAS) is a rare and underdiagnosed disease that suggests a common genetic substrate between certain clusters. In patients with an autoimmune disease, an attentive search for other autoimmune pathologies is necessary.

Keywords: *multiple autoimmune syndrome, vitiligo, biermer ulcerativecolitis, Hashimoto, lupus Sjogren*



Figure 1. *Hunter's glossitis.*
B12 Hypovitaminosis signs: Hunter's glossitis characterized by atrophy of lingual papillae.



Figure 2. *Poriasiform subacute lupus.*
Non-infiltrated erythematous-squamous lesions (called psoriasiforms) on photoexposed areas on the forearms.

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[Abstract:0493]

RITUXIMAB AS MAINTENANCE THERAPY FOR ANCA ASSOCIATED VASCULITIS WITH RECURRENT DISEASE

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Objective: Evaluate the response to maintenance treatment with Rituximab, in patients with systemic ANCA-Associated Vasculitis with a recurrent evolutionary course after multiple immunosuppressive treatments.

Methods: Retrospective, observational study, that includes a subgroup of patients with ANCA associates vasculitis treated with Rituximab, with an induction regimen of 1 g on days 0 and 14, associated with and immunosuppressant treatment.

At month 6, patients received a maintenance regimen with 2 doses of 500 mg separated by two weeks and 500 mg every six months. The evolutionary course is evaluated assessing clinical improvement and modification in ANCA levels.

Results: 9 patients were included (7 women, 78%), mean age 53 years. Diagnosed with microscopic polyangiitis 4 (45%), granulomatosis with polyangiitis 4 (45%) and eosinophilic granulomatosis with polyangiitis 1 (10%). The average number of immunosuppressants previously used was 2.5 per patient, 7 (78%) were already treated with rituximab.

Patients have received a median of 25 months of Rituximab maintenance treatment. All but one patient (8/9, 89%) experienced sustained clinical improvement. Among the responders, complete remission of renal (2, 23%) and respiratory (6, 67%) involvement was achieved, and incomplete remission of ENT involvement (3, 33%). The joint symptoms were controlled in 80% of patients (4/5). The patient who did not improve had predominant ENT symptoms.

Seven patients (78%) were ANCA positive, 5 of them experienced a reduction in titer (72%). Among patients who were already treated with rituximab, only 1 did not improve (14%).

Keywords: *rituximab, ANCA-associated vasculitis, maintenance therapy*

[Abstract:0494]

ANTI-GLOMERULAR BASEMENT MEMBRANE DISEASE: A CASE REPORT

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Antiglomerular basement membrane disease is a rare vasculitis. It presents as glomerulonephritis and/or alveolar hemorrhage. Despite treatment, recovery of renal function after initiation of hemodialysis is rare.

A 65-year-old woman presented to the emergency department with vomiting, diarrhea, and decreased urine output. Blood analysis revealed acute renal failure with hyperkalemia, metabolic acidemia, and elevation of inflammatory markers. Bilateral pulmonary infiltrates and no abdominal complications were noted on thoracobadomino-pelvic tomography. Acute bacterial gastroenteritis was suspected and treated with ceftriaxone and azithromycin. This treatment was continued for 7 and 5 days, respectively. Microbiological results were negative. The renal dysfunction was refractory to fluid therapy and later to diuretic therapy. On day 2, hemodialysis was required. Later tests included a positive anti-nuclear antibody, negative anti-DNA and extractable nuclear antigen, positive anti-MBG and negative anti-neutrophil cytoplasmic antibody. She was diagnosed with anti-glomerular basement membrane disease involving the kidneys and lungs. She was started on plasmapheresis and immunosuppressive therapy (corticosteroids and cyclophosphamide). There was a decrease in antibody titers without renal recovery after 16 sessions of plasmapheresis. Due to nosocomial infection with SARS-CoV-2 and pancytopenia, cyclophosphamide was discontinued (after 25 days). She was discharged on dialysis. Corticosteroids were stopped after 4 months of treatment with a negative result for antibodies.

It should be noted that a bacterial infection may have been the trigger for the immunologic state. Even when the etiopathology is known, this case illustrates the need to maintain a high index of clinical suspicion.

Keywords: *kidney failure, immunosuppression*

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[Abstract:0501]

ANTI-LGI1 AUTOIMMUNE ENCEPHALITIS IN A PATIENT WITH RHEUMATOID ARTHRITIS AND MGUS

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Anti-leucine-rich glioma inactivated 1 limbic encephalitis (anti-LGI1 LE) is one of the most frequent autoimmune encephalitides, commonly coexisting with other autoimmune diseases, although a causal relationship between such entities has not been established yet. Herein, we present the case of anti-LGI1 LE in a 71-year-old, male patient with rheumatoid arthritis (RA) and Monoclonal Gammopathy of Unknown Significance (MGUS). The development of anti-LGI1 LE in a patient already suffering from RA or MGUS - let alone both - has never been reported hitherto to our knowledge, despite the fact that both diseases have been associated with the development of autoimmune phenomena.

The patient was admitted due to an epileptic seizure and disorientation during the past few days. He was firstly treated as suffering from viral encephalitis based on altered mental status and increased number of CSF lymphocytes in conjunction with normal blood tests. After hospital exit, patient's symptoms relapsed, so he was re-admitted to the hospital and a thorough and extensive workup was initiated. The identification of anti-LGI1 autoantibodies both in the patient's serum and CSF, in combination with the typical deteriorating psychomotor symptoms that had emerged, led to the diagnosis of anti-LGI1 LE. Notably, the patient also exhibited persistent hyponatremia, a common feature of anti-LGI1 LE. The initial treatment with high dose corticosteroids and IVIG led to symptoms remission. Interestingly enough, the patient experienced a mild relapse one month later, so rituximab was added to the treatment regimen, resulting in complete regression of disease symptoms.

Keywords: *autoimmune encephalitis, rheumatoid arthritis, MGUS, hyponatremia*

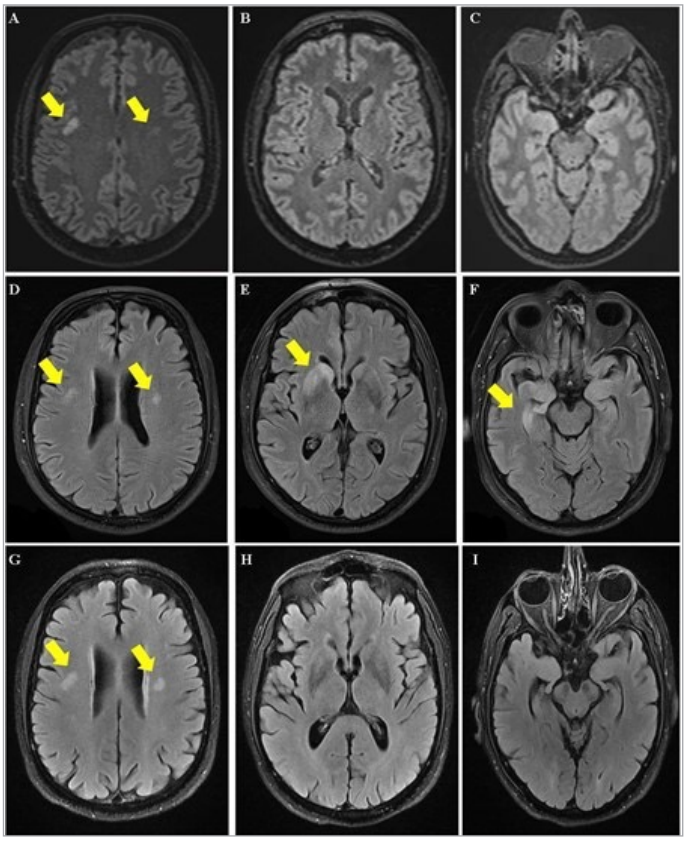


Figure 1. *Brain FLAIR-weighted MRI imaging showing the progression and remission of lesions caused by anti-LGI1 limbic encephalitis in a 71-year old male patient with rheumatoid arthritis and MGUS.*
Figures A-C are upon initial presentation. Figures D-F are upon first readmission and initial diagnosis of anti-LGI1 LE. Figures G-I are 10 days after the initial treatment with intravenous (IV) corticosteroids and IV immunoglobulin. Figures A, D and G show the initial-old lesion bilaterally. Figures B, E and H show the presence [E] and remission [H] of inflammatory lesions in the medial temporal lobes (next to the frontal horns of the lateral ventricles), predominantly in the right one. Figures C, F and I show the presence [F] and remission [I] of inflammatory lesions in the medial temporal lobes at the level of basal ganglia between the caudate lobe and the lentiform nucleus.

Test		Result
	Imaging	
Brain MRI scan		2 new foci with enhanced signal in T2, FLAIR and DWI sequences and low signal in T1 sequence in the medial temporal lobes
Chest and abdomen CT scan		Normal
Whole-body PET-CT scan		Normal
	Electrophysiological Studies	
EEG		Normal
	Laboratory Studies	
Antibodies for autoimmune and/or paraneoplastic encephalitis (CSF and serum)		[+] anti-LGI1 in serum and CSF [+] anti-VGKC in serum
Tau, p-Tau and 14-3-3 protein levels in CSF		Normal
Anti-ganglioside antibodies in CSF		Negative
CSF oligoclonal bands		[+] oligoclonal bands type 5 (indicative of antibody production in the periphery and passive transition in the CSF)
Various serum autoantibodies		Only [+] anti-CCP
	Neuropsychological evaluation (upon readmission)	
MMSE		24/30 (Orientation: 5/10, Registration: 3/3, Attention/Calculation: 5/5, Recall: 1/3, Language: 8/8, Copying: 1/1)
MoCA		16/30 (Visuospatial/Executive: 3/5, Naming: 3/3, Attention: 5/6, Language: 0/3, Abstraction: 2/2, Delayed recall: 0/5, Orientation: 2/6)
FAB		8/18 (Similarities: 2/3, Lexical fluency: 0/3, Motor series: 3/3, Conflicting instructions: 0/3, Go-No Go: 0/3, Grasp reflex: 3/3)

Table 1. *Workup performed during readmission for the differential diagnosis of encephalopathy.*
MRI: Magnetic Resonance Imaging; FLAIR: Fluid-attenuated inversion recovery; DWI: Diffusion-weighted imaging; CT:

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Computerized Tomography; PET: Positron Emission Tomography; EEG: Electroencephalogram; CSF: Cerebrospinal fluid; LGI1: leucine-rich glioma inactivated 1 protein; VGKC: voltage-gated potassium channel; CCP: cyclic citrullinated peptide; MMSE: Mini Mental State Examination; MoCA: Montreal Cognitive Assessment; FAB: Frontal Assessment Battery.

[Abstract:0503]

PARTICULARITIES OF ADULT ONSET STILL'S DISEASE IN A TUNISIAN CENTRE

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Introduction: Adult onset Still's disease is a rare auto-inflammatory systemic disease. Its clinical manifestations are diverse. The aim of our study is to describe the epidemiological, clinical, therapeutic and evolutionary characteristics of adult onset Still's disease in the Tunisian centre.

Methods: This is a retrospective and descriptive study of patients with Still's disease from 2010 to July 2023.

Results: 25 patients were included with a mean age of 48.3 years [18-73 years]. The sex ratio (M/F) was 0.25. The mean age of diagnosis was 40 years [16-58 years]. Inaugural clinical manifestations included fever (n=25), inflammatory arthralgias (n=23), odynophagia (n=17), and deterioration of general condition (n=9). The clinical examination revealed a skin rash (n=19), arthritis (n=11) involving the wrists (n=5), knees (n=5), and elbows (n=1), superficial lymphadenopathy (n=8) in the cervical (n=4) and axillary (n=3) regions, hepatomegaly (n=5), and splenomegaly (n=6). A biological inflammatory syndrome was observed in all cases, characterized by an accelerated ESR (mean 71.3 mm), elevated CRP (mean: 172 mg/l), an inflammatory profile on serum protein electrophoresis, hyperleukocytosis, and hyperferritinemia (mean 8349 µg/l). Glycosylated fraction was tested in 8 patients returning <20% in 7 cases. Hepatic cytolysis (n=9) with elevated LDH (n=13) was observed. Radiologically, abdominal ultrasound was performed (n=16), revealing hepatomegaly (n=6), splenomegaly (n=6), and hepatic steatosis(n=5). All patients were treated with corticosteroids at a dose of 1mg/kg/day (n=21) and 0.5 mg/kg/day (n=4). Six patients received methotrexate treatment. The outcome was favorable under treatment in the majority of patients (n=22).

Conclusions: Adult-onset Still's disease is a rare autoimmune disease characterized by heterogeneity in clinical and paraclinical manifestations. A better understanding of this pathology is essential to guide strategies for early diagnosis and appropriate treatment.

Keywords: *adult-onset Still's disease, autoimmune disease, rare*

[Abstract:0505]

THE INFLAMMATORY PROFILE FOR ADULT-ONSET STILL'S DISEASE

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Introduction: Adult onset Still's disease is a rare auto-inflammatory systemic disease causing a biological inflammatory syndrome. The aim of our study is to describe the inflammatory biological profile of adult-onset Still's disease.

Patients and Methods: This is a retrospective and descriptive study of patients with Still's disease from 2010 to July 2023.

Results: 25 patients were included with a mean age of 48.3 years [18-73 years]. The sex ratio (M/F) was 0.25. The mean age of diagnosis was 40 years [16-58 years]. Inaugural clinical manifestations included fever (n=25), inflammatory arthralgias (n=23), odynophagia (n=17), and a deterioration of general condition (n=9). A biological Inflammatory Syndrome was observed in all patients, characterized by an accelerated ESR (mean 71.3mm), elevated CRP (mean 172 mg/l), an inflammatory profile on serum protein electrophoresis (n=25) with elevated alpha-2 globulin in 23 cases (mean 11.7 g/l,[8-20.5 g/l]), and hyper gamma globulin in 17 cases (mean 15.2 g/l, [7.1-21.5 g/l]). Hyperleukocytosis was present in all patients [9000-31500/mm] and hyperferritinemia (mean: 8349 µg/l). Glycosylated fraction was tested in 8 patients, returning <20% in 7 cases.

Infection and immunological tests were negative in all patients. Treatment consisted of corticosteroids in all cases, combined with methotrexate(n=6), resulting in favorable biological improvement marked by the resolution of the biological inflammatory syndrome within a mean period of 3 weeks [10- 45 days].

Conclusions: Adult-onset Still's disease is a rare autoimmune disease characterized by heterogeneity in clinical and paraclinical manifestations. The study of the inflammatory profile is crucial for both diagnosis and monitoring the disease's progression. Investigating other inflammatory markers, especially pro-inflammatory cytokines, may represent a promising therapeutic approach for treatment.

Keywords: *adult-onset Still's disease, autoimmune disease, Inflammatory profile*

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[Abstract:0525]

A FIRST-TIME OBSERVATION OF SILICONE-ASSOCIATED SCLERODERMA-LIKE SYNDROME: HOW TO RECOGNIZE AND DIAGNOSE SIMILAR CASES

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While silicone breast implants are generally considered safe medical devices, implant ruptures and silicone leakage are not uncommon. In this case report, we describe a unique case of a woman with ruptured breast implants and longstanding unexplained systemic symptoms. She developed an atypical and localized scleroderma-like syndrome, with a first-time observation of histopathologically confirmed silicone depositions in distant sclerotic cutaneous biopsies. This observation with silicone-specific modified oil-red-o (MORO) staining strongly suggests a direct inflammatory reaction to the distant (sub) cutaneous silicone depositions from her ruptured breast implants. We propose clinical recommendations on how to recognize and diagnose possible similar cases.

Keywords: *silicone breast implants, scleroderma, silicone leakage*

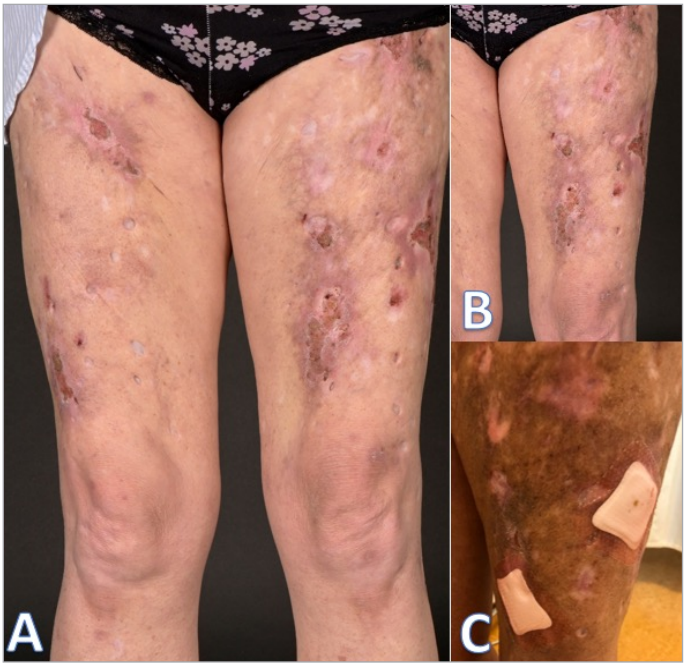


Figure 1. *A. Abscesses and sclerosis, upper legs, September 2020. B. Left Upper Leg before UV-A light therapy, September 2020. C. Left upper leg after UV-A light therapy, March 2021.*

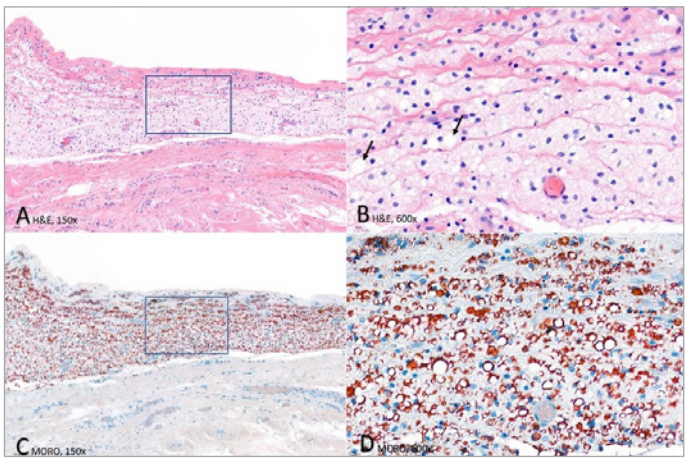


Figure 2. *Capsule of the left breast. A: H&E, 150x. Upper half of the picture showing the capsule with at the top the surface, extensive accumulation of foamy macrophages in the capsule and the lower half showing surrounding connective tissue. B: H&E staining, 600x. High magnification of the marked square in 2A with glassy vacuoles in macrophages (some indicated with an arrow). C: modified oil-red-o (MORO) staining, 150x. Low magnification. D: high magnification of the MORO staining showing strong positive staining of especially the periphery of the vacuoles.*

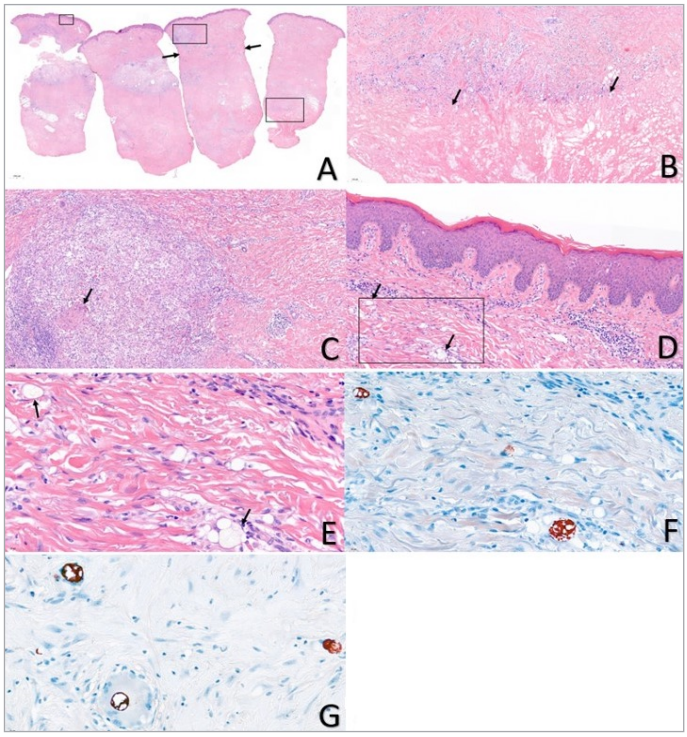


Figure 3. *Skin excision right upper leg. A: H&E staining, 10x. 4 slices of skin and subcutaneous tissue with at the top the epidermis with extensive underlying fibrosis reaching into the deep subcutaneous tissue (the arrows indicate the margin between the dermis and subcutaneous tissue) with only some islands of fat tissue remaining. B: H&E staining, 100x. Magnification of the most right marked square in 3A which shows vital tissue at the top and necrotic tissue at the bottom (arrows indicate the transition from vital to necrotic tissue). C: H&E staining, 100x. Magnification of the middle marked square in 3A with a granulomatous inflammation of the dermis with an included island*

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of squamous epithelium (arrow). D: H&E staining, 200x. Magnification of the most left marked square in 3A with perivascular lymphocytic infiltrate in the dermis and some foamy macrophages (arrows). E: H&E staining, 550x. High magnification of the marked square in 3D with the foamy macrophages (arrows) with intracytoplasmic glassy material. F: MORO staining, 550x. The same area as shown in 3E with strong positive staining of the foamy macrophages. G: MORO staining 500x. An area deep in the subcutaneous tissue also showing strong positive staining in macrophages.

[Abstract:0528]

PD-1 INDUCED AUTOIMMUNE HEPATITIS - A CASE REPORT

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Introduction: Autoimmune hepatitis (AIH) is a chronic hepatitis of unknown etiology, but the presumed trigger may involve medications. PD-1/PD-L1 (Programmed death-ligand 1 Inhibitor) are a group of immune checkpoint inhibitors (ICIs) widely used as front-line treatment of multiple types of cancer. It has substantially improved the prognosis of many advanced malignancies. However, with the use of ICIs, more and more unique spectrum of side effects had been identified including immune-related adverse events.

Case Presentation: 68-years-old male, history of urothelial carcinoma with lung and liver metastasis, under treatment with pembrolizumab. Admitted in Internal Medicine ward due to progressive liver enzymes elevation consistent with hepatocellular pattern (ALT and AST up to 1000+U/L). Diagnostic work up was done that were all inconclusive, including autoimmune markers, viral serology. He agreed to undergo liver biopsy which suggested interface hepatitis. His clinical condition was complicated with hepatic encephalopathy. By exclusion of all other possible etiologies, PD1 induced AIH was considered and started prednisolone 1 mg/Kg/day. He gradually improved with liver enzymes downtrending progressively to normal. Corticosteroids were tapered down weekly and patient was discharged and referred for follow up in Internal Medicine consultation. At present, his clinical condition is significantly improved, and his liver function completely recovered.

Conclusions: In the era of emerging use of PD-1 inhibitors, autoimmune adverse events will be encountered more frequently. Although rare, PD-1 induced AIH should be kept in mind after exclusion of other causes. Therefore, we would like to present this case to increase awareness for this condition.

Keywords: *autoimmune hepatitis, PD1 - inhibitor, immune adverse events*

[Abstract:0551]

ORO-FACIAL INVOLVEMENT IN SYSTEMIC SCLEROSIS

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Introduction: Systemic sclerosis (SS) is an autoimmune disease characterized by the triad of inflammation, fibrosis, and vasculopathy with collagen deposition in connective tissue. Orofacial involvement is not spared and can constitute significant functional and aesthetic discomfort. The aim of this study is to describe the oro-facial manifestations in patients with SS.

Patients and Methods: This is a retrospective descriptive study that includes patients diagnosed with SS according to the ACR/EULAR 2013 criteria and followed in a Internal Medicine Department from June 2011 to July 2023.

Results: We collected data from 51 patients with an average age of 54.8 years [26-89 years]. The gender ratio (M/F) was 0.2, with 10 males and 41 females. The mean age at the time of systemic sclerosis (SS) diagnosis was 49 years [21-88 years]. Among the 51 patients, 45 had oro-facial involvement, resulting in a prevalence of 88.2%.

Oro-facial examination revealed the presence of cutaneous sclerosis (82%), microstomia (80%), exaggeration of perioral radial folds (46.6%), facial telangiectasias (31%), skin pigmentation disorders with areas of hyperpigmentation and depigmentation (13%), and mandibular and alveolar bone erosion (8%) leading to tooth loss.

Xerostomia was reported by 20 patients, including 12 patients with associated Sjogren's syndrome.

Conclusions: Orofacial manifestations are far from uncommon in systemic sclerosis (SS) but are often overlooked by patients and underdiagnosed by clinicians. The aesthetic and functional impact can sometimes be significant, leading to disability.

Keywords: *systemic sclerosis, orofacial, autoimmune disease*

[Abstract:0554]

A NEW ETIOLOGY OF VASCULAR PURPURA: ORAL ANTICOAGULANTS

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Introduction: Vascular purpura is a dermatological condition characterized by non-blanching hemorrhagic lesions upon diascopy. The etiologies of vascular purpuras are diverse. We report two cases of vascular purpura induced by direct oral anticoagulants (DOACs).

Case Presentation: Case 1: An 82-year-old female, hypertensive

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and diabetic, was admitted for the management of deep vein thrombosis in the left external iliac vein secondary to prolonged bed rest. The patient was treated with rivaroxaban. Three weeks later, there was the onset of vascular purpura on both lower limbs without signs of severity. Skin biopsy revealed a histological pattern of lymphocytic vasculitis with negative immunofluorescence. A drug-induced origin was considered and the evolution was marked by improvement after discontinuation of rivaroxaban.

Case 2: A 73-year-old patient, recently diagnosed with stage T3N3M1b bronchopulmonary neoplasia, was not under any treatment, presented with a complicated right femoropopliteal and leg thrombosis. The patient was treated with apixaban. Three weeks later, the patient reconsulted for vascular purpura without signs of severity. Skin biopsy revealed a morphological pattern of purpura secondary to a hemostatic disorder. Immunofluorescence was negative. A drug-induced origin was considered, and the approach involved discontinuing apixaban and transitioning the patient to enoxaparin, resulting in a good clinical outcome: improvement of purpuric lesions after stopping the DOACs.

Conclusions: Vascular purpura associated with DOACs represents novel etiology to be added to the spectrum of vascular purpura etiologies. The underlying pathophysiological mechanism remains incompletely elucidated, with some authors suggesting a potential association with hemostatic disturbances. Discontinuation of DOACs often leads to a significant improvement in cutaneous lesions.

Keywords: *vascular purpura, oral anticoagulants, novel etiology*

[Abstract:0557]

HYPOPHOSPHATEMIC OSTEOMALACIA CAUSED BY INTRAVENOUS IRON THERAPY

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Case Description: We present the case of a 63-year-old male diagnosed of hereditary haemorrhagic telangiectasia and iron-deficiency anaemia due to chronic bleeding from gastrointestinal telangiectasias secondary to his underlying condition. The patient required treatment with weekly blood transfusions and intravenous ferric carboxymaltose every two weeks.

He complained of bilateral mechanical thoracic pain not related to prior trauma, with fluctuating intensity but severe enough to require opioids for relief. Blood tests revealed phosphate levels ranging from 1.25 to 2.5 mg/dL, calcium between 8.83 and 9.06 mg/dL, parathormone 79 pg/mL, 25-hydroxy vitamin D 7 ng/ml, and alkaline phosphatase 138 IU/L. Additionally, a bone scan with HDP–Tc99m showed rib fractures along with other nonspecific findings suggesting alterations in the phosphocalcic metabolism.

Clinical Hypothesis: Hence, secondary osteomalacia due to hypophosphatemia, likely related to repeated administration of ferric carboxymaltose, was considered the most probable cause of the clinical picture. Oral phosphate and vitamin D treatment were initiated, resulting in clinical and biochemical improvement. Regarding gastrointestinal bleeding, bevacizumab was administered, leading to discontinuation of transfusion dependency and parenteral iron therapy.

Discussion and Learning Points: Hypophosphatemia is a common adverse effect of intravenous iron administration, often asymptomatic. Severe cases can lead to hypophosphatemic osteomalacia, observed in association with various iron formulations, although most cases have been linked to ferric carboxymaltose.

Keywords: *osteomalacia, ferric carboxymaltose, hypophosphatemia*

[Abstract:0560]

LIPODISTROPHY IN DERMATOMYOSITIS

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Objectives: Show the rare relationship between dermatomyositis and lipodystrophy, especially in the acquired form in adults.

Materials and Methods: A series of 2 cases of dermatomyositis with acquired lipodystrophy in adults evaluated in our service.

Results: The first case is a 43-year-old woman with dermatomyositis, with muscle and skin involvement and calcinosis, who was treated until she was 15 years old with prednisone and azathioprine, and is evaluated in 2022 for a new outbreak, which is treated with metrotexate and prednisone. The examination showed the attached images showing lipodystrophy.

The second case is a 47-year-old woman with dermatomyositis, corticosteroid dependent, treated sequentially with metrotexate, mycophenolate, rituximab, again metrotexate, mycophenolate, tocilizumab, mycophenolate and finally, immunoglobulins and tofacitinib who is in the present. Upon examination, the attached images showed lipodystrophy.

Discussion: Lipodystrophy is a rare disease that is usually associated with infections, metabolic disorders or autoimmune diseases. In the case of dermatomyositis, it mainly affects young people (10-40% of them), its appearance in adults being exceptional. Although it can present as 3 phenotypes (generalized, partial or focal), it is usually partial in its extent. The etiopathogenesis is unknown although it is related to those dermatomysitis with a prolonged and severe course or those that present with calcinosis. Unlike acquired juvenile lipodystrophy, it has not been seen to be related to metabolic syndrome or insulin resistance.

Conclusions: In conclusion, lipodystrophy in patients with

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dermatomyositis usually appears at a young age; in adults, it is related to the course and prolonged and serious treatments.

Keywords: *lipodistrophy, dermatomyositis, rare disease*

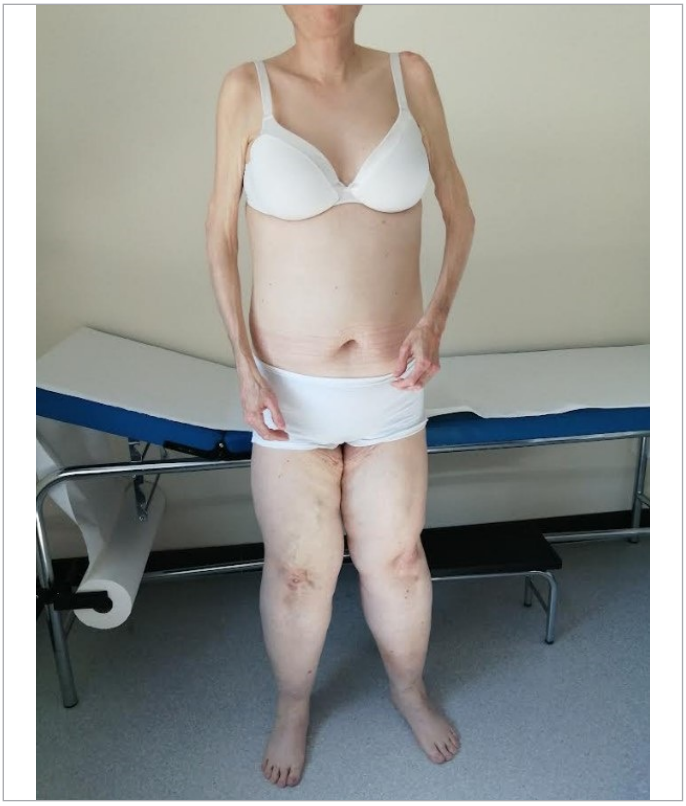


Figure 1. *Case 1.*

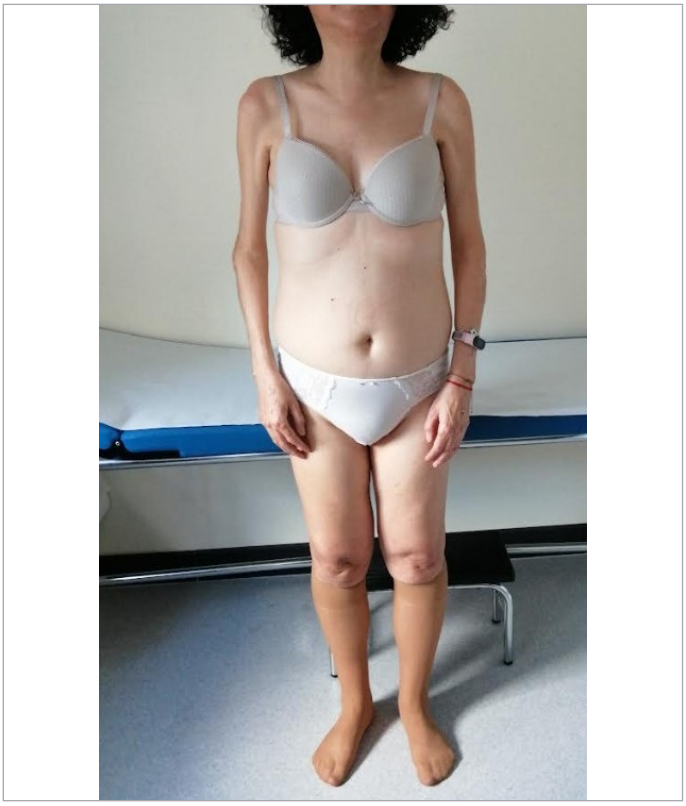


Figure 2. *Case 2.*

	Pac. 1	Pac. 2
Edad	43	47
Edad diagnóstico	8	18
Ac. asociado	Tif-1	Tif-1
CPK	484	569
Glucosa	92	75
AST	-	34
ALT	28	42
GGT	13	28
Fosf. Alc.	54	61
Colesterol total	170	254
HDL	52	75
LDL	103	144
Triglicéridos	74	174
C3	133	151
Ecografía abdominal (hígado graso)	-	normal

Table 1.

[Abstract:0566]

RELATIONSHIP BETWEEN CANCER AND SARCOIDOSIS IN TERTIARY HOSPITAL

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Introduction and Objectives: The relationship between sarcoidosis and cancer is known, having described a higher incidence of cancer in patients with sarcoidosis and the development of sarcoidosis in patients previously diagnosed and treated for cancer. We studied the relationship between both in a cohort of patients with sarcoidosis followed in our center.

Materials and Methods: Descriptive, retrospective study in patients followed up for sarcoidosis in outpatient clinics. The information was obtained from the review of medical records of patients under follow-up by sarcoidosis in the Systemic Autoimmune Diseases Unit between January 2019 and December 2022.

Results: Among the 63 patients with sarcoidosis (mean age 58 years), 36% were men. 9 patients had associated cancer (14.2% with sarcoidosis), either as a history or during its evolution, with 5 men (55%) with a mean age of 63.8 years. Except for one, the tumors were described with prior to the diagnosis of sarcoidosis with a median of 5.5 years. In most of the patients the diagnosis of sarcoidosis was made casually during cancer follow-up and they were asymptomatic. Only one patient was being treated for sarcoidosis.

Conclusions: Cancer occurs in an older population and predominantly in men. In our cohort there is a wide variety of tumors and in the majority cancer preceded the diagnosis of sarcoidosis. Sarcoidosis was discovered incidentally as part of the tumor monitoring in most cases. The most common involvement is lymphadenopathy mediastinal, followed by pulmonary and

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cutaneous. Its prognosis is generally favorable. Generally requiring treatment

Keywords: *sarcoidosis, cancer, relationship, incidence*

[Abstract:0567]

PREVALENCE AND CO-PREVALENCE OF IMMUNOGENETIC FACTORS IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES (IIM) IN A THIRD LEVEL OF COMPLEXITY

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Purpose: Autoantibodies and HLA alleles have been found to correlate with clinical phenotypes in patients with IIM across different regions globally. Our study investigates the immunogenic characteristics and its co-prevalence with clinical features in a specific Colombian population.

Methods: Cross-sectional study conducted on patients diagnosed with IIM. The panel of fifteen autoantibodies specific to myositis and those associated with it. The HLA DQA1, DQB1, DR alleles were determined using real-time polymerase chain reaction technique.

Results: A total of 68 patients were included in the study, with a mean age of 52.2 years and predominantly female (73%). The most common myopathy was dermatomyositis (DM) (58%), followed by polymyositis (PM) (25%), antisynthetase syndrome and immune-mediated necrotizing myositis (5.8%) each and inclusion body myositis in 3 cases. Table 1 shows other clinical features. Autoantibodies were studied in 23 (33%) patients and HLA in 20 (50%); within the non antisynthetase Mi-2 was found (52%) related to DM (83%), skin manifestations Gottron (75%) and Heliotrope (58%) and HLA DQB1*03 (50%), Jo-1 antisynthetase antibody (30%) relating to PM (85%), pulmonary involvement (14%) and HLA DQB1*03 (28%); Ku associated antibodies (13%) relating to DM (66%), Gottron and Heliotrope (66%) pulmonary involvement (33%) and HLA-DQB1*03 (33%).

Conclusions: This study is the first of its kind in Colombia, analyzing HLA haplotypes and myositis autoantibodies in IIM patients. The findings revealed varying co-prevalences between clinical features and the immunogenic profile, providing insight into the characteristics of the local population and supporting further research with larger sample sizes.

Keywords: *dermatomyositis, polymyositis, autoantibodies, HLA, myositis*

Myopathy Characteristics	DM 40 (58.8)	PM 17 (25)	ASyS 4 (5.8)	IMNM 4 (5.8)	IBM 3 (4.41)
Gender Women Men	32 (80) 8 (20)	11 (64) 6 (35)	4 (100) 0	1 (25) 3 (75)	2 (66) 1 (33)
Age at diagnostic (Mean)	41	56	59	49	73
Muscle weakness (MRC median) Proximal Distal	3 4	3 4	4 5	4 4	3 4
Cancer Breast	1 (2.5) 1 (100)	1 (5.8) 1 (100)	1 (25) 1 (100)	0 0	0 0
ILD NSIP UIP AIP	5 (10) 2 (5) 1 (2.5) 2 (5)	4 (23) 3 (17.6) 0 1 (5.8)	3 (75) 1 (25) 2 (50) 0	0 0 0	0 0 0
Enzymes (mean) IU/L CPK Aldolase	6500 55	3816 19	2656 27	12197 108	5944 110
Clinical features Gottron's Heliotrope Ulceration Calcinosis Raynaud's Arthritis Dysphagia Myocardial Oral ulcers	29 (72) 28 (70) 2 (5) 2 (5) 4 (10) 12 (30) 23 (57) 0 1 (2.5)	1 (5.8) 1 (5.8) 0 0 0 3 (17.6) 7 (41) 0 0	2 (50) 2 (50) 2 (50) 0 1 (25) 1 (25) 2 (50) 0 0	0 0 0 0 0 0 2 (50) 1 (25) 0	1 (33) 1 (33) 0 0 0 0 2 (66) 1 (33) 0
Autoantibody Jo1 Mi-2 TIF1g MDA5 NXP2 SAE1 Ku PM- Scl 100 PM- Scl 75 SRP PL-7 PL-12 EJ OJ Ro-52 None	18 (40) 1 (5.5) 10 (55.5) 1 (5.5) 1 (5.5) 0 (0) 1 (5.5) 2 (11.1) 0 0 0 0 1 (5.5) 0 0 0 2 (11)	14 (82) 6 (42) 1 (7.14) 0 0 0 0 1 (7.14) 0 0 0 1 (7.14) 1 (7.14) 0 0 0 0 4 (28.5)	3 (75) 0 1 (33) 0 0 0 0 0 0 0 0 2 (66) 0 0 0 0 0	2 (50) 0 0 0 0 0 0 0 0 0 0 0 0 1 (25) 0 0 (0) 1 (25)	0 (0) - - - - - - - - - - - - - - - -
HLA alleles	11 (27)	5 (29)	3 (75)	1 (25)	0 (0)
HLA-DQA1 03:01 05:01 02:01 04:01 01:01,02:02,03:03	2 (18) 0 0 1 (50) 1 (50) 0	1 (20) 0 0 0 0 1 (100)	2 (66) 0 1 (50) 0 0 1 (50)	0 0 0 0 0 0	- - - - - -
HLA-DQB1 03 0401/0402 02 05 06:01 06:02-8 03:02 or 03:03 03:01 or 03:04 03:05 or 04:01/04:02 05:01 05:03 05:02/05:04	11 (100) 8 (72) 3 (27) 5 (45) 1 (9) 0 0 2 (18) 1 (9) 0 0 0 0	5 (100) 2 (40) 4 (80) 0 0 0 1 (20) 1 (20) 0 1 (20) 0 0 0	3 (100) 3 (100) 0 0 1 (33) 0 1 (33) 2 (66) 1 (33) 0 0 0 0	1 (100) 0 1 (100) 1 (100) 0 0 0 0 0 0 0 0 0	- - - - - - - - - - - - -
HLA-DR 01 03 04 08 07 09 10 11 12 13 14 15 16	11(100) 7 (63) 1 (9) 5 (45) 3 (27) 6 (54) 0 0 0 0 2 (18) 1 (9) 0 0	5 (100) 1 (20) 0 2 (40) 4 (80) 1 (20) 0 0 0 0 1 (20) 1 (20) 0 0	3 (100) 0 0 2 (66) 0 0 0 0 0 0 1 (33) 0 0 1 (33)	1 (100) 1 (100) 1 (100) 0 0 1 (100) 0 0 0 0 0 0 0 0	- - - - - - - - - - - - - -

Table 1. *Characteristics according to myopathy.*
DM, Dermatomyositis; PM, Polymyositis; ASyS, antisynthetase syndrome; IMNM, immune-mediated necrotizing myopathy; IBM, inclusion body myopathy; ILD, interstitial lung disease; CPK, creatine phosphokinase; NSIP, Nonspecific Interstitial Pneumonia; UIP, usual interstitial pneumonia; AIP, acute interstitial pneumonia.

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[Abstract:0571]

INTERSTITIAL LUNG DISEASE ASSOCIATED WITH SYSTEMIC SCLEROSIS AND INFLAMMATORY MYOPATHIES: 2-YEAR FOLLOW-UP OF A COHORT OF PATIENTS TREATED WITH CYCLOPHOSPHAMIDE AND METHYLPREDNISOLONE PULSES

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Purpose: To evaluate the efficacy and safety of the combination cyclophosphamide-methylprednisolone (CYC-MP) as induction therapy for interstitial lung disease (ILD) associated with systemic sclerosis (SSc) and inflammatory myopathies (IM).

Methods: Observational study of routine clinical care data of 38 patients with ILD associated to SSc or IM, diagnosed between 2013 and 2021 and treated with intravenous CYC-MP followed by a reduced prednisone plus immunosuppressive regime. CT scan changes, functional parameters (FVC and DLCO) and clinical evolution (mMRC scale) were collected at 12 and 24 months.

Results: Thirty-eight patients (73.7% women, mean age 61.4 years) were included: 20 (52.6%) SSc and 18 (47.3%) IM. SSc patients received a lower maximum dose of prednisone (5.3 mg/day vs 9 mg/day, p=0.006). Stability or improvement in FVC after 24 months was observed in 80.5% of patients, with no significant differences between groups (77.7% SSc vs 83.3% IM, p=1.00). 86.1% of patients had better or stable mMRC scale scores after 24 months (72.2% SSc vs 100% MI, p=0.023). The CT scan and the DLCO improved or remained stable in 86.1% and 80.5% patients, without significant differences between groups. 6 patients (2 MI, 4 SSc) suffered an infection requiring hospitalization. No SSc renal crisis were observed. Two patients suffering SSc died due to progression of the disease.

Conclusions: Intravenous CYC-MP, followed by maintenance therapy with low-dose prednisone and immunosuppressive drugs, had a favourable effect on the progression of SSc and IM-associated ILD with a low occurrence of adverse events and a reduced load of oral glucocorticoids.

Keywords: *interstitial lung disease, systemic sclerosis, inflammatory myopathies, cyclophosphamide, methylprednisolone pulses.*

VARIABLES	Systemic sclerosis	Inflammatory myopathies	p value
Mean FVC at 12 months	86 (72-95)	83.8 (67-104)	0.693
Mean FVC at 24 months	86.5 (71-94)	89.5 (70-102)	0.748
Mean FVC evolution	-1.33 (11.94)	+7.17 (13.04)	0.093
Mean DLCO at 12 months	66.5 (49-77)	58.5 (51-66)	0.327
Mean DLCO at 24 months	61.5 (49-80)	58.5 (51-66)	0.762
Mean DLCO evolution	-1.59 (12.36)	+4.23 (11.73)	0.119
FVC stability or improvement at 12 months	14 (70%)	15 (83.3%)	0.334
FVC stability or improvement at 24 months	14 (77.7%)	15 (83.3%)	1.000
DLCO stability or improvement at 12 months	16 (80%)	15 (83.3%)	0.791
DLCO stability or improvement at 24 months	14 (77.7%)	15 (83.3%)	1.000
mMRC stability or improvement at 12 months	15 (75%)	17 (94.4%)	0.101
mMRC stability or improvement at 24 months	13 (72.2%)	18 (100%)	0.023
CT scan stability or improvement at 12 months	14 (70%)	16 (88.9%)	0.154
CT scan stability or improvement at 24 months	16 (88.8%)	15 (83.3%)	1.000

Table 1. *Results. Evolution of CT scan, functional and clinical parameters at 12 and 24 months.*

[Abstract:0573]

ERTAPENEM INDUCED DRUG RELATED EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS) SYNDROME - A CASE REPORT

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Case Description: An 89 year-old female, with kidney impairment (sCr: 3.0 mg/dL) due to autosomal dominant polycystic kidney disease, is admitted with sepsis with bacteremia from an infected kidney cyst.

Treatment with ertapenem was initiated and blood and urine cultures later isolated ertapenem-sensitive *E. coli*. A 30-day course was implemented due to slow clinical progression along with the assumption of infected cysts.

On the first day after completion, an extensive dorsal pruriginous maculopapular rash associated with labial edema developed which motivated treatment with systemic corticosteroids. After 3 days, the facial edema worsened, and the rash had progressed to the chest, abdomen and limbs (~55% body surface area).

She remained apyretic and no lymphadenopathies were documented.

Clinical Hypothesis: Ertapenem associated DRESS syndrome.

Diagnostic Pathways: A complete blood count showed eosinophilia (max 1100/µL) and IgE was elevated (786 UI/mL). Viral, *mycoplasma* and *chlamydia* serologies and immune studies were unremarkable. There was no internal organ involvement. A skin biopsy revealed perivascular lymphocytic infiltrate with presence of eosinophils, compatible with DRESS. Due to clinical deterioration, corticosteroid dose was increased, and a 5-day course of IgIV was initiated. The rash progressively improved with desquamation, and on the 25th day of corticosteroid treatment, it was practically resolved and allowed tapering. These characteristics allow classification as a "possible DRESS syndrome".

Discussion and Learning Points: One should always consider DRESS syndrome as a possible diagnosis when a patient presents

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with a typical rash after 2-8 weeks of treatment of a new drug even in the absence of other clinical hallmarks.

Keywords: *ertapenem, DRESS syndrome, eosinophilia*

[Abstract:0578]

NEVER FORGET THE (DREADED AND DEVASTATING) DIFFUSE ALVEOLAR HEMORRHAGE

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Introduction: Diffuse Alveolar Hemorrhage (DAH) is a rare and life-threatening complication of lupus systemic erythematous (SLE).A high index of suspicion is required for diagnosis to initiate a rapid treatment. Even so the diagnostic delay can be up to 4 days. DAH evolves in hours-few days and is the syndrome associated with SLE with the highest mortality.

Clinical Case: Female, 72 years. History of SLE with episodes of DAH in the past. Presenting to be clinically stable from SLE, on prednisolone at a minimal dose. She went to the emergency department for prostration, hemoptoic expectoration and hematuria. The patient denied dyspnea, without respiratory insufficiency. Analytically with anaemia, lymphopenia and CRP elevation. X-ray with cottony infiltrates in the right lung field, superimposed on previous exams.

A) Favorable evolution in the first 48 hours. Subsequently, with a rapid clinical deterioration requiring oxygen. A chest scan was performed, which confirmed a new episode of DAH, requiring intubation in the following hours, and was hospitalized in intensive care-

B) In intensive care, she received corticosteroids, immunoglobulins and plasmapheresis with a favorable evolution of DAH after 7 days, progressively without hemoptoic expectoration, she was extubated and transitioned to oxygen therapy. During the weaning from oxygen therapy, she developed a septic condition, with an unfavorable outcome.

Conclusions: DAH is defined by the sudden onset of dyspnea, hemoptysis, hypoxemia, tachycardia, cough and abrupt drop in Hb. However, diagnosis depends on a high clinical suspicion, sometimes non-existent due to the subclinical presentation. In the present case, although the clinic frustrated and the evolution was favorable in the first 48 h ours, the evolution can be rapid and septic conditions may be involved in the dysfunction, contributing to a devastating outcome. The authors intend to warn that the DAH clinic is silent in autoimmune diseases, which may delay the recognition of this entity.

Keywords: *diffuse alveolar hemorrhage,s ystemic lupus erythematosus, life-threatening*

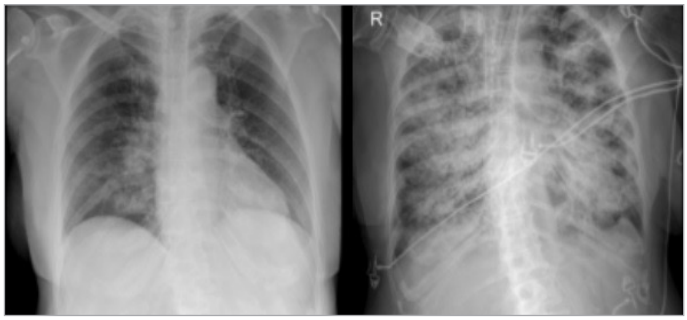


Figure 1. *Postero-anterior chest radiograph upon admission (left) and approximately 10 days later (right).*

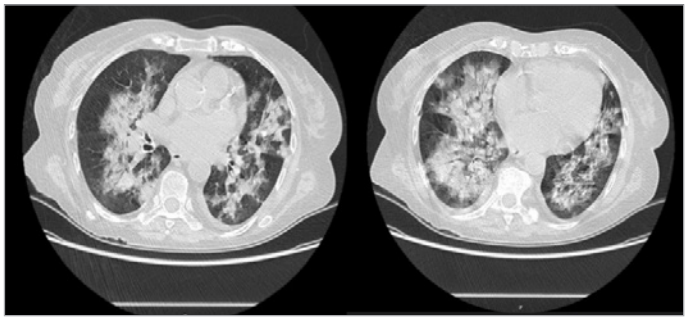


Figure 2. *CT scan sections about 1 week after admission.*

[Abstract:0606]

GOOD SYNDROME: WHAT IS IT REALLY? GOOD OR BAD

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Introduction: Patients with immunodeficiency are prone to bacterial, fungal, viral and opportunistic infections. In this case report, we present a patient with Good's syndrome, which we detected in a patient with a positive COVID-19 test and opportunistic infection a long time after a thymoma operation.

Case: A 70-year-old woman with history of asthma and chronic inactive hepatitis B presented with long lasting abdominal pain and diarrhea, with multiple previous admissions. According to lab tests no infectious cause was detected for diarrhea. During colonoscopy, caecal ulcers were detected, methylprednisolone and mesalazine was started with suspicion of Crohn's disease. The patient, whose symptoms persisted under this treatment, was admitted to internal medicine clinic for further tests. Upon

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developing of cough and detection of lymphopenia in blood tests, COVID-19 swab test was performed and came as positive. In addition, after white plaques reminiscent of fungal plaques that cause dysphagia were detected inside mouth. CMV tests performed on blood and biopsy samples from colonoscopy were substantially high. During the follow-up, 7 different Covid-19 samples taken over course of 40 days all came as positive. The patient who had fungal, CMV, and long-lasting COVID infections was suspected of having an immunodeficiency disorder. Upon further tests, immunoglobulin levels were found to be low and the patient who had thymectomy 16 years ago and clinic of immunodeficiency was diagnosed with Good's syndrome and intravenous immunoglobulin treatment was started.

Conclusions: Good's Syndrome is a late-onset primary immunodeficiency disorder consisting of thymoma, hypogammaglobulinemia and immunodeficiency triad. Immunodeficiency syndromes should be considered for diagnosis in patients with long-term COVID infection and opportunistic infection.

Keywords: *Good syndrome, hypogammaglobulinemia, long COVID, thymoma*

[Abstract:0618]

TAKAYASU'S ARTERITIS AND CROHN'S DISEASE: A RARE ASSOCIATION ?

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In 2019, a 25-year-old woman, attended the emergency department for holocranial headache, weakness on the left side of the body and dysarthria.

Given the initial suspicion of stroke, cerebral CT angiography was performed, with no obstruction of large vessels. Intravenous fibrillation was decided with slight improvement of the language disorder. The following day, due to persistent neurological focality, cerebral CTA was repeated. Internal carotid dissection and inflammation of large caliber arteries were found. Vasculitic carotid dissection was suspected, possibly Takayasu's disease, so treatment was started with glucocorticoids and methotrexate with excellent and rapid response, remaining in clinical remission.

After one year, she began with an increase in the number of stools, of soft consistency and hematochezia. He starts a digestive study with a final diagnosis of Crohn's disease. Discussed in a multidisciplinary session Rheumatology-Digestive System, it was decided to modify the therapeutic scheme associating infliximab, maintaining MTX at a moderate dose and glucocorticoids at the minimum effective dose. Excellent response to treatment, remaining in clinical remission.

The association between IBD and AT is rare, but the existence of common pathophysiologic pathways indicates that these conditions may be strongly associated. Since both diseases can be treated with the same agents, a delay in diagnosis may occur. We believe that knowledge of this association, although infrequent, is important from a clinical point of view, so a thorough physical examination should be performed in patients with CD to detect signs of its association with AT in order to treat its possible complications early.

Keywords: *Takayasu's arteritis, Crohn's disease, vasculitis*

[Abstract:0624]

PEDIATRIC VACCINATIONS AND MYOSITIS: A CASE REPORT

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We present the case of a 3-year-old girl, diagnosed with polyarticular form of JIA ANA+/FR-. After two years of treatment with methotrexate and remaining in clinical remission, a precautionary suspension of treatment was decided. The following year he received the vaccinations corresponding to the pediatric calendar: meningococcus and DTaP (tetanus, diphtheria and pertussis) and in the following weeks he began with fever and intense myalgia with elevated creatinine kinase (1292 U/l) and fever of 37.3ºC. After performing electromyogram and muscle biopsy, the diagnosis of inflammatory myopathy dermatomyositis pattern is confirmed, responding excellently to treatment with glucocorticoids and methotrexate.

There is no strong evidence to support the relationship between vaccination with different pediatric vaccines and the development of first episodes or recurrences of inflammatory myopathies. Vaccination and revaccination with inactivated vaccines is considered safe in children with a medical history of Guillain-Barré (GBS), Bell's palsy, optic neuritis (ON), myasthenia gravis (MG), chronic inflammatory demyelinating polyneuropathy (CIDP) and immune-mediated inflammatory myopathy (IM). Caution is suggested when vaccinating against influenza, tetravalent meningococcal conjugate vaccine (MCV4) and pneumococcal disease and avoiding immunization with tetanus toxoid in patients with CIDP. Patients with immune-mediated acquired inflammatory neuromuscular disorders should be vaccinated with live vaccines before initiating immunosuppressive therapy.

Keywords: *inflammatory myopathies, myositis, vaccination*

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[Abstract:0638]

THERAPEUTIC MODALITIES AND CLINICAL OUTCOMES IN AN ADULT COHORT WITH LRBA DEFICIENCY AND CTLA4 INSUFFICIENCY

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Background: LPS-responsive beige-like anchor (LRBA) deficiency (LRBA-/-) and cytotoxic T-lymphocyte-associated antigen-4 (CTLA4) insufficiency (CTLA4+/-) are mechanistically overlapped diseases presenting with recurrent infections and autoimmunity. The effectiveness of different treatment regimens remains unknown.

Objective: To determine the comparative efficacy and long-term outcome of therapy with immunosuppressants, CTLA4-immunoglobulin (abatacept), and hematopoietic-stem-cell-transplantation (HSCT) in a single-country multicenter cohort of 49 patients with a 5-year median follow-up.

Methods: The 49 patients (24 LRBA-/- and 25 CTLA4+/-) were followed and evaluated at baseline and every 6 months for clinical manifestations and response to the respective therapies.

Results: The LRBA-/- patients exhibited a more severe disease course than did the CTLA4+/- patients, requiring more immunosuppressants, abatacept, and HSCT to control their symptoms. Among the 28 patients who received abatacept as either a primary or rescue therapy, sustained complete control was achieved in 17 (60.7%) without severe side effects. In contrast, most patients who received immunosuppressants as primary therapy (n = 31) showed either partial or no disease control (77.4%), necessitating additional immunosuppressants, abatacept, or transplantation. Patients with partial or no response to abatacept (n = 11) had longer disease activity before abatacept therapy, with higher organ involvement and poorer disease outcomes than those with a complete response. HSCT was performed in 7 LRBA-/- patients; 6 patients (85.7%) showed complete remission, and 1 patient awaits second transplantation. HSCT and abatacept therapy gave rise to similar probabilities of survival.

Conclusions: Abatacept is superior to immunosuppressants in controlling disease manifestations over the long term, especially when started early, and it may provide a safe and effective therapeutic alternative to transplantation.

Keywords: *inborn errors of immunity, LPS-responsive beige-like anchor, abatacept, cytotoxic T-lymphocyte-associated antigen-4, hematopoietic stem cell transplantation, immune dysregulation*

[Abstract:0646]

A CLINICALLY AMYOPATHIC DERMATOMYOSITIS DIAGNOSIS IN A MALE PATIENT OUTSIDE A TYPICAL PRESENTATION

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The broad family of idiopathic inflammatory myopathies (IIMs) includes dermatomyositis (DM), polymyositis, myositis as part of a rheumatic disease overlap syndrome, myositis of the anti-synthetase syndrome, immune-mediated necrotizing myopathy, and inclusion body myositis (IBM). Autoimmune-inflammatory myopathies are characterized by muscle weakness, positive autoantibodies in the blood, and inflammatory infiltrates in muscle tissue.

Anti-MDA-5 antibody was first reported in a Japanese population of CADM patients. Since then, it has been identified that patients with anti-MDA-5 autoantibodies show more severe cutaneous findings and rapidly progressive interstitial lung disease than patients without anti-MDA-5 autoantibodies. Our patient had no hand swelling, arthritis, arthralgia, muscle pain, and diffuse hair loss, which are more common in the reports of patients with anti-MDA-5 antibodies. With normal muscle strength, our patient was considered to have the least features of skeletal findings of DM.

In conclusion, this case represents a male patient with CADM who did not show much of typical clinical and laboratory findings reported so far. CADM has been reported markedly more frequent in women. Whether male patients tend to show less specific clinical and laboratory findings is unknown, but our patient shares similarities with a previous male patient from the US who also lacked several typical manifestations. On the other hand, the current report emphasizes the critical role of even minor skin changes and pulmonary involvement in the diagnosis of DM before serology testing.

Keywords: *myopathies, interstitial lung disease, anti-MDA-5 antibody*

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[Abstract:0648]

RENAL INTRICACIES IN ANKYLOSING SPONDYLITIS: A SPOTLIGHT ON AMYLOIDOSIS

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Summary: Renal involvement in ankylosing spondylitis (AS) represents a rare complication of the disease, primarily attributed to the emergence of renal amyloidosis as a secondary response to the chronic inflammatory status. The biological therapy and its effects, along with issues concerning survival, pose challenges that demand heightened attention in patients with chronic autoinflammatory diseases.

Purpose: Presentation of a case involving secondary renal amyloidosis in a patient diagnosed with ankylosing spondylitis.

Methods: We present the case of a 45-year-old male diagnosed with HLA-B27 positive AS. Since 2006, he has been managed with etanercept, a competitive inhibitor of the TNF-alpha receptor, exhibiting favorable outcomes in the context of articular manifestations. Nevertheless, from 2008 onwards, the patient has presented with azotemic retention syndrome and nephrotic-range proteinuria (>5 g/24 hours). A comprehensive diagnostic evaluation has excluded diabetes mellitus, infectious etiologies, cardiovascular factors (renal artery stenosis) and renal neoplasms. Given the chronic autoinflammatory nature of the condition, secondary renal amyloidosis has been suspected and renal biopsy revealed positive amyloid deposits upon Congo red staining. The progression of renal failure has been gradual but exacerbated, reaching an estimated glomerular filtration rate (eGFR)<15 ml/min/1.73 m² in 2022, necessitating the initiation of dialysis while maintaining etanercept therapy.

Findings: Renal amyloidosis, although rare in AS, underscores the importance of remission in the underlying disease. In studies, etanercept is shown to improve the renal function in secondary amyloidosis by reducing proteinuria. In this case, the effect of etanercept has demonstrated a mitigating effect on the rapid decline of renal function.

Keywords: *ankylosing spondylitis, AA amyloidosis, renal involvement*

[Abstract:0655]

A CASE REPORT OF POST COVID-19 NEW-ONSET DERMATOMYOSITIS

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Purpose: Various new-onset autoimmune diseases have been correlated with SARS-CoV-2 infection. We present a case of post-Covid-19 dermatomyositis in an unvaccinated patient.

Methods: A 66-year-old man, unvaccinated against SARS-CoV-2, with a history of coronary artery disease, hypothyroidism, dyslipidemia and severe COVID-19 infection 8 months ago, presented to the emergency department with a 4-month history of muscle weakness. On physical examination he had symmetrical proximal muscle weakness, dry bibasilar crackles, diffuse macularopapular rash of trunk and extremities, violaceous eruption on the periorbital skin, erythema of the shoulder and neck, Gottron's papules and hyperkeratosis and fissuring of the palms and lateral fingers. Laboratory testing showed elevated levels of muscle enzymes (CPK: 5115 U/I, AST: 124 U/I, ALT: 120 U/I, aldolase: 60.5 U/I) and LDH (809 U/I) while screening for hepatitis, neoplasmatic markers, paraproteinemia were negative. Thyroid function was normal.

Findings: Further laboratory investigation revealed the presence of ANA, anti-ENA, anti-Ro52 and anti-PL-7 autoantibodies. The electromyogram showed myopathic-type lesions in the proximal muscles. Gastrointestinal endoscopy and abdominal CT scan were negative for malignancy, while chest CT scan revealed bilateral bronchopulmonary infiltrates. Spirometry revealed restrictive lung disease. During hospitalization, the patient's rash evolved to erythroderma with itching and fever. Muscle and skin biopsies were consistent with dermatomyositis. Systemic glucocorticoids and rituximab were initiated with a favorable response.

Conclusions: The correlation between COVID-19 and dermatomyositis needs further investigation, and clinicians should be aware of the occurence of typical dermatomyositis in unusual cases of post-COVID 19 inflammatory myopathy.

Keywords: *desmatomyositis, post-COVID 19, inflammatory myopathy*



Figure 1. *Erythroderma.*

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Figure 2. Mechanic hands.

[Abstract:0662]

OSTEITIS CONDENSAS ILII: AN OVERLOOKED CAUSE OF CHRONIC LOW BACK PAIN IN REPRODUCTIVE AGE WOMEN

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Background: Osteitis condensans ilii (OCI) is the sclerotic change of the iliac bone at the side of the sacroiliac joint, commonly seen in young women after pregnancy and the puerperal period leading to chronic low back pain (CLBP). Here, we report two patients presenting with chronic low back pain and diagnosed with OCI.
Case Presentation: Two female patients with long-term CLBP started after pregnancy are presented. Both patients were evaluated for spondyloarthritis (SpA) and no inflammation signs on laboratory tests or genetic tendency were found. Straight leg raise, FABER, and FADIR tests were negative bilaterally. Modified Schober's test was in normal range, the fingertip-to-floor distance was zero, chest expansion, sacroiliac compression, cervical rotation, lumbar lateral flexion, occiput-to-wall, and tragus-to-wall tests were normal, also. Radiographs and magnetic resonance imaging (MRI) confirmed the diagnosis of OCI and physical therapy in combination with non-steroidal anti-inflammatory drugs and myorelaxants showed apparent benefit.
Conclusions: The two main reasons for CLBP are SpA and lumbar hernia with the need for a specific treatment approach according to inflammatory and systemic involvement of SpA and surgical approach of lumbar hernia, respectively. OCI is another reason with a frequency of 1-2.5%, especially in women of childbearing age. It is generally asymptomatic and diagnosed incidentally or missed and detected with a delay. Because chronic low back pain results in severe healthcare costs, loss of workforce, increased disability and morbidity, differential diagnosis from inflammatory and other mechanical causes is important for both early and effective treatment and management of patients.

Keywords: *osteitis condensans ilii, chronic low back pain, spondyloarthritis, pregnancy, reproductive age*

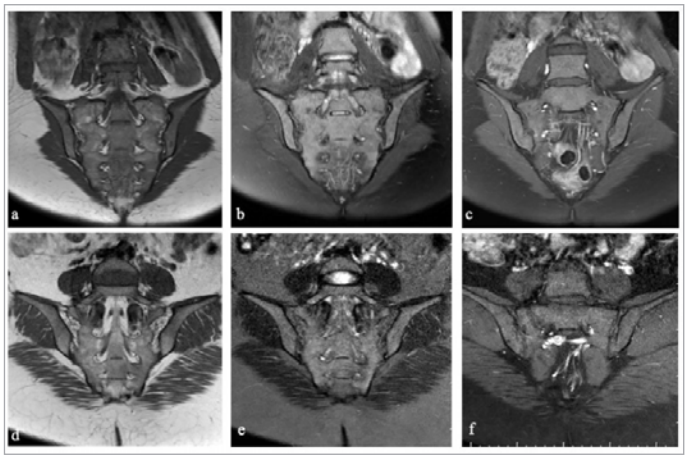


Figure 1. MRI findings of Case 1 and Case 2, respectively. In T1 (a and d) and fat-suppressed T2 (b and e) weighted images, no irregularity was observed in the sacroiliac joint, and there is a hypointense appearance compatible with subcortical sclerosis on the bilateral iliac faces, more prominent on the left. (c and f) Contrast-enhanced T1-weighted images showed lack of contrast enhancement and suggested sclerosis. In addition, no signal enhancement addressing sacroiliitis was detected in T2 and contrast enhanced T1-weighted series.

[Abstract:0677]

DESCRIOTION OF THE TREATMENT AND EVOLUTION OF PATIENTS DIAGNOSED WITH STILL'S DISEASE IN A TERTIARY CARE HOSPITAL

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Objectives: To describe the diagnostic procedures, treatments used and evolution of patients in follow-up for Still's disease in a third level hospital until 2023.
Materials and Methods: This was a retrospective descriptive case series. We included non-pediatric patients in outpatient internal medicine consultations at our center (tertiary hospital) between 1997 and May 2023 with a diagnosis of Still's disease. Clinical Documentation and the digital history of the Andalusian Public Health System. The analysis was performed with IBM-SPSS®.
Results: 14 patients were identified. The treatments used

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were NSAIDs, steroids, corticosteroid sparing and monoclonal antibodies, in most patients in combination (Figure 1). No patient controlled the disease with NSAIDs alone, 2 were controlled only with corticosteroids. The mean starting dose of corticosteroids was 30.2 mg (range 10-80). Most required stepwise treatment adding corticosteroid sparing (methotrexate 7 patients, 50%; azathioprine 2 patients, 14%; cyclosporine 6 patients, 43%; and mycophenolate 1 patient, 7%) and/or monoclonal antibodies (etanercept 7%, tocilizumab 36%, anakinra 43%). 8 patients required a switch to another monoclonal antibody during their treatment.

The disease was monocyclic in 5 patients, polycyclic in 4, and chronic in 3. Complications included hemophagocytic syndrome in 2 patient, pulmonary complications in 1, osteoporosis in 1, infection due to immunosuppression in 1. No fractures due to osteoporosis, disseminated intravascular coagulation or steroid diabetes were observed. Currently, 35.7% have active disease despite combination therapy, 28.6% have controlled disease, 28.6% are in remission and in 7.1% the current status is unknown.

Keywords: *autoimmune, treatment, Still*

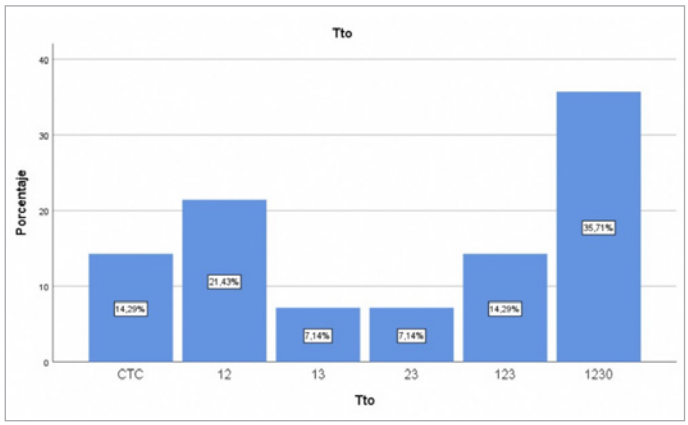


Figure 1. Corticosteroids (CTC); corticosteroids and corticosteroid sparing (12); corticosteroids and monoclonal antibodies (13); corticosteroid sparing and monoclonal antibodies (23); corticosteroids, corticosteroid sparing and monoclonal antibodies (123); NSAIDs, corticosteroids, corticosteroid-sparing and monoclonal antibodies (1230).

[Abstract:0696]

OLD DRUGS ARE STILL EFFECTIVE AND USEFUL

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Gout is a disease known since ancient times. It is due to tissue deposition of uric acid crystals. It may be asymptomatic or with episodic arthritis. In addition, it may be accompanied by tophi. Uric acid deposits trigger an inflammatory response that leads to episodes of arthritis and can be very painful and disabling. Treatment consists on drugs that reduce the synthesis, allopurinol or febuxostat, or that increase its excretion, such as probenecid or benzbromarone. For acute gout crisis, NSAIDs, colchicine, corticosteroids or, in refractory cases, IL1 inhibitors such as anakinra or canakinumab are used.

We present a case with disabling tophaceous gout which treatment is quite challenging. This is an 84-year-old woman. In 2014 she had an acute attack in the 1st MTF of the right foot, with ulcerated tophi, that kept her confined to a wheelchair. In 2008 she presented with DRESS syndrome, related to treatment with allopurinol, and in January 2014 he was prescribed febuxostat, which was discontinued due to mucocutaneous involvement. She did not tolerate colchicine. She was maintained on low doses of steroids. In March 2014, benzbromarone was started. Oral bicarbonate was associated. She had an excellent evolution with resolution of arthritis, healing of ulcerations, and a decrease in uricemia levels from 12.5 mg/dL to 6 mg/dL. After eight years of surveillance, the patient has low levels of uric acid and she is able to walk.

This case illustrates that knowledge of the entire therapeutic options is crucial in the treatment of hyperuricemia and gout.

Keywords: *gout, tophi, benzbromarone*

[Abstract:0735]

CLINICAL PROFILE AND RELEVANCE OF ANTI-NOR90 ANTIBODIES: EXPERIENCE IN A SYSTEMIC AUTOIMMUNE DISEASE UNIT

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Introduction: Anti-NOR90 antibodies have been documented in various autoimmune diseases. The objective of our study is to delve into the most clinically relevant aspects of these autoantibodies in patients within our Unit.

Materials and Methods: A single-center descriptive observational study was conducted. We reviewed patients with positive anti-NOR90 antibodies over the past 10 years, gathering clinical variables from their digital medical records.

Results: We identified 8 patients with a median age of 48 years, with the majority (7 out of 8) being women. 50% met criteria for Sjogren's syndrome, 50% for systemic sclerosis, and 25% for systemic lupus erythematosus. Two cases of systemic sclerosis were diffuse, one without scleroderma, another pre-scleroderma, and one overlapping syndrome. The most prevalent symptoms

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were dryness, cutaneous-joint involvement, and interstitial pneumonia (50%, with one of them rapidly progressive). Capillaroscopy was nonspecific in 37.5%. Most patients showed coexistence with other antibodies. Only one patient developed neoplasia during the follow-up.

Conclusions: Anti-NOR90 antibodies are of low prevalence and remain relatively unexplored. Within our cohort, we observed a notable prevalence of interstitial pneumonia, aligning with existing scientific literature. Screening for interstitial lung disease in patients with positive anti-NOR90 antibodies would be advisable. The prevalence of neoplasms was low, in contrast to other studies. Multicenter studies are necessary to better characterize these findings.

Keywords: *anti-NOR90, systemic sclerosis, autoimmune diseases*

[Abstract:0743]

ORGAN DAMAGE IN SYSTEMIC LUPUS ERYTHEMATOSUS. A PENDING TASK

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Background and Aims: To describe the baseline characteristics and the organ damage in a series of patients with diagnosis of systemic lupus erythematosus (SLE).

Methods: Retrospective descriptive study of patients diagnosed with SLE by SLICC (2012) and/or EULAR/ACR (2019) criteria in follow-up by the Systemic Autoimmune Diseases (SAD) Unit between 2018 and 2022 in a third level hospital in Madrid.

The SLICC/ACR Damage Index (SDI) was used to calculate the cumulative organ damage.

Results: Forty-six patients were included, 91% were female, 63% Caucasian and 28% Latin American. 65% of patients were receiving corticosteroid therapy.

Only 74% were on hydroxychloroquine and 47.8% of patients were receiving a corticosteroid-sparing treatment. The majority of the patients had musculoskeletal (25/41), cutaneous (22/41), cardiovascular (17/41), renal (11/41), ocular (7/41), peripheral vascular system (7/41) and neuropsychiatric damage (4/41). The mean SDI was calculated according to the years of disease evolution (1-5; 6-10; 11-15 and >15 years) obtaining an SDI of 2.11 (SD +/- 1.32); 2.60 (SD +/- 1.50); 3.00 (SD +/- 3.03) and 3.14 (SD +/- 1.86), respectively.

Conclusions: Most patients had musculoskeletal, cutaneous, cardiovascular and renal damage. The value of SDI seems to increase as the disease evolves.

Sixty-five percent of patients were receiving corticosteroid treatment and only 74% were being treated with hydroxychloroquine and 47.8% with a corticosteroid sparing agent, which is associated with greater damage. It is essential to determine early the cumulative organ damage in patients with SLE in order to try to mitigate it.

Keywords: *organ damage, SDI, SLE*

[Abstract:0744]

14-3-3H PROTEINS AS A DIAGNOSTIC MARKER, DISEASE ACTIVATION INDICATOR, AND LYMPHOMA PREDICTOR IN PATIENTS WITH PRIMARY SJOGREN SYNDROME

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Background: Primary Sjogren Syndrome (PSS) is a chronic, autoimmune, and lymphoproliferative disease of the connective tissue. In patients with PSS, the risk of developing B-cell non-Hodgkin lymphoma (NHL) increases dramatically, with a prevalence of approximately 5%. The 14-3-3 protein isoforms are phospho-serin/phospho-threonine binding proteins associated with many malignant diseases. This study aimed to evaluate the relationship between disease activity parameters and markers predicting lymphoma development in patients with PSS and 14-3-3H proteins.

Keywords: *14-3-3H protein, lymphoma, primary Sjogren syndrome*

[Abstract:0751]

POLISEROSITIS IN SYSTEMIC LUPUS ERYTHEMATOSUS - PSEUDO PSEUDO MEIGS' SYNDROME

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Background: Systemic lupus erythematosus (SLE) is an autoimmune disease with multiple clinical manifestations, such as serositis.

Purpose: We herein present the case of a 46-years-old female, diagnosed with SLE in 2006, with a long history of inconsistent medical treatment and monitorization, who was admitted to our clinic in March 2022.

Methods: At admission the patient presented with dyspnea, abdominal pain, weight loss, nausea and vomiting. Clinically, malar rash, livedo reticularis, friable hair, enlarged abdomen with build-up fluid and bilateral absence of breath sounds, were found.

Findings: Further investigations showed positive direct Coombs test with secondary iron deficiency anemia, important inflammatory syndrome, hypocomplementemia, low seric proteins, and proteinuria (1.5 g/24h). Immunology findings showed positive

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antinuclear, anti-double stranded DNA, and anti-Smith antibodies in association with high serum values of CA-125 tumoral marker. Imaging proof of polyserositis was found and pleural, pericardiac and abdominal fluid was analysed. There were no tumoral findings on computed tomography. Despite immunosuppressant and high dose cortisone treatment, renal impairment persisted. Renal biopsy was practiced and mezangioprolipherative glomerulonephritis stage II was discovered. Although several immunosuppressants were used, the disease remained highly active, until treatment was switched to cyclophosphamide with slow, but favourable evolution.

Conclusions: A case of persistent polyserositis, non-responsive to various immunosuppressants. We considered the diagnosis of pseudo-pseudo Meigs' syndrome, with few cases reported in literature, in a patient with SLE, polyserositis (ascitis, pleural effusion), high serum levels of CA-125, but with no tumoral evidence. Important to underscore the relationship between CA-125 inflammation and disease acitvity.

Keywords: *polyserositis, systemic lupus erythematosus, CA-125*

[Abstract:0760]

TOCILIZUMAB IN BEHÇET'S DISEASE: A MULTICENTER STUDY OF 30 PATIENTS

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Objective: To evaluate tocilizumab (TCZ) efficacy in patients with refractory Behçet's disease (BD).

Methods: This is a multicenter study of 30 patients fulfilling the international criteria for BD and treated with TCZ at different European referral centers. The clinical response was evaluated at 6 months from TCZ initiation.

Results: Ninety percent of patients with BD were refractory or intolerant to anti–tumor necrosis factor (anti-TNF) agents. Overall, TCZ was effective in 25 (83%) patients with BD of whom 18 (60%) and 7 (23%) were complete and partial responders, respectively. The complete response was 67%, 60%, and 42% in patients with uveitis (18/30), neurological manifestations (5/30), and mucocutaneous and/or articular (7/30) manifestations, respectively. TCZ had a significant steroid-sparing effect allowing patients to decrease their median daily prednisone dose from 20 (IQR 10-40) mg/day to 9 (IQR 5-13) mg at 6 months ($P < 0.001$). The number of patients with BD needing concomitant disease-modifying antirheumatic drug therapy fell from 7 (23%) to 4 (13%) at 6 months.

Mild to moderate side effects were observed in 6 (20%) patients, and 3 (10%) presented with serious adverse events (pneumonia, intestinal perforation, and septicemia) requiring therapy discontinuation in 2 cases.

Conclusions: TCZ seems to be an effective alternative to anti-TNF agents in treating BD-related uveitis and neurological manifestations.

Keywords: *Behçet's disease, neuro-Behçet, tocilizumab, uveitis*

[Abstract:0834]

CHECKPOINT INHIBITORS, A MILESTONE. BUT WHAT ABOUT COMPLICATIONS?

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Case: We present a case of a 52-year-old male with lung adenocarcinoma undergoing Pemetrexed-Pembrolizumab treatment. Hospitalized for a worsening month-lasting diarrhea, with blood, mucus, and elevated inflammatory markers, the performed proctoscopy revealed immune-mediated colitis (IMC) superinfected with cytomegalovirus (CVM). Valganciclovir and corticosteroids are started with no appropriate response so vedolizumab is associated but given a worsening in symptoms is then switched to infliximab, along with pembrolizumab cessation, achieving a complete resolution of symptoms.

Discussion: The immune system's role in recognizing and eliminating tumours through T-cell recognition of tumour-associated antigens is crucial. Immune checkpoints, like CTLA-4 and PD-1 that naturally inhibit immune response, are sometimes exploited by tumours, inhibiting T-cell activation. Immune checkpoint inhibitors (ICIs) enhance cancer treatment but may induce dysregulated responses, causing, among others,

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gastrointestinal diseases, including IMC, an entity resembling inflammatory bowel disease (IBD). IMC and IBD share clinical and diagnostic similarities, being IMC classified into four stages. ICI discontinuation is not indicated in stage I, transiently in stage II during symptomatic treatment, and permanently in stage IV; remaining stage III management still on debate. Treatment involves intravenous corticosteroids, and if unresponsive, anti-TNFα like Infliximab is indicated. Vedolizumab is considered for anti-TNFα refractory IMC. Overall, understanding these immune dynamics informs effective cancer treatment while managing associated complications.

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1.Som A, Mandaliya R, Alsaadi D, Farshidpour M, Charabaty A, Malhotra N, Mattar MC. Immune checkpoint inhibitor-induced colitis: A comprehensive review. World J Clin Cases. 2019 Feb 26;7(4):405-418.

Keywords: *checkpoint inhibitors, inmune mediated colitis, cancer treatment*

[Abstract:0862]

HENOCH SCHONLEIN PURPURA WITH RENAL AND LUNG INVOLVEMENT

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Case Description: We present the case of a 54-year-old man, with a clear medical history, admitted after pruritic rash development, compatible with palpable purpura and petechiae affecting the lower extremities, combined with arthralgias involving the ankles, hands and feet. Physical examination showed a purpuric rash on the extensor surfaces of the lower extremities, as well as swollen and painful joints. The rest of the examination was normal and laboratory tests showed microscopic hematuria. Initial imaging tests showed a mosaic pulmonary attenuation on CT imaging.
Diagnostic Pathways: Further laboratory tests (serum C3, C4, CANCA, pANCA, anti-MPO, anti-PR3, anti-GBM, serum protein electrophoresis, cryoglobulins), gastroscopy, were negative or normal. Due to strong clinical suspicion of Henoch- Schonlein angiitis, we proceeded to skin and kidney biopsy which showed small vessel leycocytoclastic vasculitis with IgA immune deposits as well as glomerular mesangial deposition of igA complexes.
Conclusion and Discussion: The patient was started on corticosteroid therapy with rapid improvement of arthralgias.
It is worth noting that pulmonary involvement has been rarely described in patients with Henoch- Schonlein purpura, occurring more often in adults and commonly manifesting as diffuse alveolar haemorrhage and occasionally as usual interstitial pneumonia or interstitial fibrosis.

Keywords: *henoch schonlein purpura, renal involvement, pulmonary involvement, complications*



Figure 1. *Pruritic rash development.*

[Abstract:0873]

A RARE CASE OF FAMILIAL COLD AUTOINFLAMMATORY SYNDROME 2 (FCAS2) RECOGNIZED IN ADULT LIFE

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Purpose: FCAS2 is an autosomal dominant disorder that belongs to the spectrum of systemic autoinflammatory diseases and sporadically affects both adults and children. It is characterized by episodic and recurrent rash, urticaria, arthralgia, myalgia, and headache, accompanied by fever and evidence of inflammation.
Methods: A 49-year-old female was admitted to our department due to a 3-week history of intermittent fever (>39°C). She was previously well, apart from recurrent headaches and flares of urticaria treated with antihistamines. No family history for systemic/autoinflammatory diseases was reported. Clinical examination revealed cervical lymphadenopathy, splenomegaly and mild hepatomegaly. Laboratory tests upon admission showed elevated erythrocyte sedimentation rate, C-reactive protein, serum amyloid-A and mild cholestasis.
Findings: Extensive investigation excluded infectious diseases, neoplasms, hematologic malignancies and systemic autoimmune disorders. The patient also underwent liver biopsy, essentially demonstrating Kupffer cell activation and neutrophil infiltration. Subsequently, genetic testing was performed, revealing the

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mutation F402L (c.1206C>G) of NLRP12 gene, that has been frequently associated with FCAS2. The patient was initially treated with colchicine and anakinra with resolution of fever. However, due to a presumably drug-induced rash, steroids were administered with clinical and laboratory response.

Conclusions: Although rare, autoinflammatory diseases should be suspected in patients with recurrent or persistent inflammation unexplained by other causes, such as infection or malignancy.

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Keywords: *autoinflammatory syndrome, familial, FCAS2*

[Abstract:0917]

50-YEAR-OLD WOMAN WITH PARAVERTEBRAL MASS

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Case Description: A 50-year-old woman consults for intermittent fever and lancinating dorsal pain. She refers malaise and three diarrheal stools per day. She has morbid obesity and Crohn's disease.

Examination and chest-X-ray normal. Spine MRI with anterior paravertebral mass D6 -D10, negative microbiological and quantiferon studies, ESR 103 and CRP 50-110 mg/L. ANA 1/160 with ELISA and negative ANCA, normal biopsy and negative histochemical and molecular study, PET with hypercaptation of intervertebral and vertebral disc with rarefaction of fat without uptake. New biopsy with chronic sclerosing inflammatory process with fibroedema and lymphomonocytic and plasmocellular infiltrate. The patient continued with fever and pain. The diagnosis of paravertebral inflammatory pseudotumor was made and azathioprine and prednisone were prescribed with improvement. The case was reconsidered by reviewing the evolution and images and a diagnosis of axial spondyloarthropathy associated with IBD with fatty changes was made.

Discussion: Crohn's disease and ulcerative colitis are the pathologies most frequently associated with axial spondyloarthropathy associated with inflammatory bowel disease with a prevalence of 1-26%. It usually causes axial involvement and inflammatory lumbar pain with stiffness. 4-16% of asymptomatic patients have sacroiliitis. MRI is indicated if there are symptoms. ESR and CRP are markers of activity. Differential diagnosis is made with septic arthritis, reactive and whipple's disease. Treatment is with exercise, NSAIDs and antiTNF drugs if pain persists.

Keywords: *fever, paravertebral mass, Inflammatory bowel disease*

[Abstract:0942]

PROGNOSTIC FACTORS RELATED WITH SURVIVAL AND MORTALITY IN SECONDARY HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

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Summary: Secondary hemophagocytic lymphohistiocytosis (sHLH) has a high mortality rate in hospitalized patients, so it is essential to evaluate prognostic markers and mortality predictors.

Purpose: To assess analytic variables obtained at diagnosis and describe prognostic markers that influence survival in patients with sHLH.

Methods: Retrospective and descriptive study, where demographic and clinical data was obtained from patients diagnosed with secondary hemophagocytic lymphohistiocytosis (sHLH) in a tertiary hospital from 2018 to 2023.

Findings: Our data showed an increase in survival in female patients (70% versus 43% in male patients). 59% of the patients with sHLH survived. There were three principal causes (Figure 1). There was a 100% survival rate in patients with an autoimmune antecedent, in contrast to infection (67%) and malignancy (33%). A worse prognosis was found in patients with hypertriglyceridemia (65%), hypertransaminasemia (60%), coagulopathy (47%), anemia, elevation of lactate dehydrogenase and ferritin. Erythrocyte sedimentation rate was directly related to a better survival rate.

Discussion: A high clinical suspicion is crucial in sHLH due to a mortality rate that ascends to 88% in many studies. In our study, 41% of patients passed away. It is important to highlight the increase in mortality in male patients and an initial increase in ferritin and a slowed decrease the days following the diagnosis, as described in previous studies.

Conclusions: Strict monitoring is necessary in patients with sHLH, especially with certain biochemical markers as described in this study. Decreased mortality in contrast to other studies, especially in patients with autoimmune diseases, is noteworthy.

Keywords: *hemophagocytic lymphohistiocytosis, autoinmunity, prognostic factors.*

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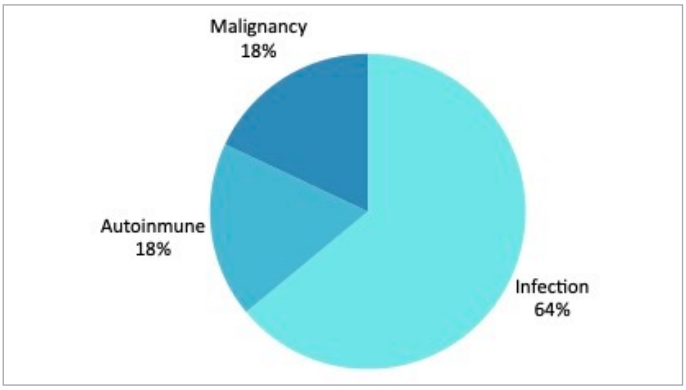


Figure 1. Causes of secondary hemophagocytic lymphohistiocytosis in our study.

[Abstract:0945]

SENSORY APHASIA IN A YOUNG PATIENT WITH NO RELEVANT HISTORY. WHAT ARE WE MISSING?

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A 36-year-old male, with no personal or epidemiological history of interest, was brought in for a 1-hour history of inability to maintain related dialectics. The previous day he consulted because he started having headache, neck pain, nausea and diarrhea. Temperature up to 37.3ºC. The physical examination revealed aphasia of expression and some nuchal rigidity. A complete analysis was performed, B12, folic acid, rotavirus and adenovirus in stool, ECG, chest x-ray, head CT, TSA Doppler, within normal limits. In CSF, hyperproteinorrachia (PT 73.7 mg/dL), lymphocytic pleocytosis (32 leukocytes, 70% mononuclear), normal glucose and ADA negative. Gram stain and culture were negative. Acyclovir 10 mg/kg/ every 8 hours was instituted, presenting complete resolution of symptoms after 48 hours. Cranial MRI, without relevant findings. In the complete serology requested upon admission, positive IgM Ab for leptospira interorgan was obtained, everything being consistent with meningeal leptospirosis. Penicillin was started at 6 million units every 6 hours, and he was discharged, with favorable evolution. Leptospirosis is usually biphasic. Symptoms usually resolve within a week; However, after 1-3 days, it may recur (immune phase) coinciding with the development of antibodies, with central nervous system (CNS) involvement being an important feature. Signs and symptoms may be intense headache, photophobia, neck stiffness and altered mental status, which may be in the absence of liver, kidney or respiratory disease. In addition to meningitis (5-24%), a variety of CNS and peripheral findings have been reported in about 7% (Table 1).

Keywords: leptospirosis, infections, neurology

Phase	Neurological complications
First	Headache, pain, meningismus, encephalopathy.
Second	Meningitis.
Third	Meningoencephalitis, encephalitis, encephalomyelitis, hemorrhage (subarachnoid, subdural, spinal). Cranial neuropathy.
SNP abnormalities	Radiculoneuritis, mononeuritis, plexitis, axonal or demyelinating neuropathy.

Table 1. Neurological complications of leptospirosis.

[Abstract:0956]

A RARE PRESENTATION OF STIFF-PERSON SYNDROME WITH A NONSPECIFIC FOCAL MYOSITIS: A CASE REPORT

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Introduction: The Stiff-Person Syndrome (SPS) is a rare disease whose incidence is estimated at approximately 1 in a million individuals in the general population. We present the case of a patient diagnosed with the idiopathic SPS.

Observation: A 36-year-old patient presented with a subacute dorsal myalgia. He reported episodes of torticollis and back stifness. He was afebrile, had hyperlordosis with a paravertebral contracture and tenderness. Neurological and cognitive examinations were normal. Laboratory investigations, including markers of muscular damage, were within normal range. Spinal MRI revealed T2 hyperintensity in the semispinalis, erector spinae, trapezius, as well as the intervertebral muscles, described as signs of myositis. The electromyogram revealed a continuous motor unit activity in agonist and antagonist muscles, suggestive of SPS. Blood culture, tuberculosis skin test, antinuclear antibodies, anti-Glutamic Acid Decarboxylase 65 (anti-GAD65) and onconeuronal antibodies were negative. Analysis of the cerebrospinal fluid, including anti-GAD65 test, was normal. Echocardiogram and thoracic-abdominal-pelvic scanner were normal. The MRI description was rather explained by the continuous contraction of the affected muscles. The patient was commenced on diazepam and baclofen.

We noticed that paravertebral contracture became less noticeable. Diagnosis of SPS was established according to Dalakas criteria. Investigations for an underlying neoplasm were normal. Associated autoimmune disorders have been ruled out. Treatment included diazepam, baclofen, intravenous immunoglobulin and corticosteroids. The symptoms responded well and the patient showed important improvement.

Conclusions: The SPS is a rare condition whose diagnosis can be delayed. Characteristic signs such as axial rigidity or muscle spasms should alert the physician.

Keywords: Stiff-Person syndrome, stiff-man syndrome, myositis, autoimmune

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[Abstract:0958]

A CASE OF DRESS SYNDROME INITIALLY TREATED AS SCARLET FEVER

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Purpose: To present the case of a patient with drug-related eosinophilia with multiorgan involvement, that was originally misdiagnosed as scarlet fever.

Methods: A 74-year-old woman presented with a diffuse rash and fever of 6 days. She had been prescribed clindamycin, because of her grandson having been diagnosed with scarlet fever a few days prior. Reported comorbidities were hypertension, chronic kidney disease and gout flare-ups, for which allopurinol had been started 4 weeks ago. On examination, a low-grade fever, an itching maculopapular rash of trunk and extremities and palm desquamation were noted. Laboratory results showed elevated inflammatory markers, abnormal liver biochemistry and deterioration of renal function. Abdominal ultrasound and X-Rays were normal.

Findings: Intravenous methylprednisolone (1 mg/kg/day) was initiated, due to high suspicion of a drug-induced hypersensitivity reaction, despite absence of eosinophilia. Blood and urine cultures, as well as serologic tests against HHV-6,7,8, EBV and CMV were negative. After admission, deterioration of liver biochemistry with concomitant rise of eosinophil count (1900cells/μL) were observed. MRCP showed no abnormal findings. High doses of intravenous methylprednisolone (1g/day for 3 days) were administered. Subsequently, eosinophil count decreased, skin lesions regressed and liver and kidney functions improved. Treatment continued with prednisolone, eventually withdrawn after 8 weeks, with normalization of liver and kidney values.

Conclusions: Most DRESS patients recover completely after drug withdrawal. A number of high-risk drugs is to blame, with allopurinol being the most common. This case report highlights the challenges in management of such a patient and supports the choice of alternative gout therapies.

Keywords: *DRESS, methylprednisolone, allopurinol, gout*

[Abstract:0985]

COEXISTENCE OF SCLEROMYOSITIS AND MYASTHENIA: A RARE ASSOCIATION!

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Introduction: The multiple autoimmune syndrome is a rare entity defined by the coexistence of at least three autoimmune diseases. We report the rare observation of an association of systemic sclerosis, antisynthetase syndrome, and myasthenia gravis. To our knowledge, this is the second reported case.

Observation: A 45-year-old female patient was referred for systemic sclerosis, without respiratory or muscular involvement. Two years later, she presented with dyspnea and muscle fatigue. She had mechanic's hands and bilateral symmetric proximal muscle weakness. Laboratory investigations revealed CK levels at 995 IU/L and positive anti-PL7 antibodies. Thoracic CT showed diffuse interstitial lung disease. Spirometry demonstrated a restrictive lung disease. The electroneuromyogram (ENMG) showed a myopathic pattern. Diagnosis of anti-synthetase syndrome was established. Treatment was initiated with monthly cyclophosphamide infusions. Seven months later, she presented with left ptosis and worsening dyspnea and muscle fatigue. An intravenous immunoglobulin infusion was administered, then the patient was commenced on mycophenolate mofetil. She showed a slight improvement, then she developed dysphagia. Brain MRI and cerebrospinal fluid were normal. Onconeuronal antibodies were negative.

The ENMG did not reveal any decrement, but the acetylcholine receptor antibody level was positive at 1.12 nmol/L. Consequently, with the presence of ptosis, dysphonia and dysphagia, the diagnosis of myasthenia gravis was established. Imaging did not reveal a thymoma. A favorable therapeutic response was achieved with intravenous immunoglobulins and pyridostigmine.

Conclusions: Inflammatory myopathies and myasthenia are two pathologies that can quickly pose a threat to the patient's life, highlighting the importance of a prompt diagnosis and therapeutic plan.

Keywords: *scleromyositis, myasthenia, antisynthetase syndrome, the multiple autoimmune syndrome*

[Abstract:0991]

AN INFLAMMATORY DISEASE OF THE AORTA

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Case Description: A 58-year-old male with no allergies, diagnosed in 2019 with syncope that is studied in internal medicine department. No pain or fever. Temporal pulses present and anodyne examination. An echocardiography revealed a thickening of the aortic wall and dilatation of 5 cm with mild insufficiency. MRI angiography showed dilatation of the aortic root and ascending aorta with a circumferential thickening of the wall. Positron emission tomography (PET) scan demonstrated inflammatory uptake in the aorta. Negative blood cultures. Laboratory tests showed an elevation of ESR to 10 and CRP of 20. Autoimmunity (ANA, ANCA), immunoglobulins and infectious serologies negative. IgG levels elevated and gG4 subclasses 500

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mg/dl. Corticosteroid therapy was started with a descending pattern first but an elevation of RFA was objectified as well as an increase in inflammatory activity in PET so rituximab was added with analytical response and improvement of inflammation.

Clinical Hypothesis: Aortitis in relation to IgG4 disease.

Discussion: IgG4 disease is a fibroinflammatory, multiorgan and immune-mediated disease that simulates other entities (infectious, inflammatory or tumor). In our case the diagnosis was made with serological and imaging criteria, as well as discarding causes of exclusion and confirming the response with corticosteroids. Rituximab is an anti CD20 drug, 2nd line for this disease and in which several clinical trials have shown to reduce relapses with low doses of corticosteroids. It has also been shown to reduce the inflammatory burden as well as the inflammatory fibrotic reaction. This case report highlights the importance of being aware of IgG4 disease in the presence of any organic inflammation in order to diagnose it and establish early treatment to avoid progression and future relapses.

Keywords: *IgG4 disease, inflammation, aortitis*

[Abstract:1002]

FIRST CASE OF IGG4-RELATED DISEASE IN CANTONAL HOSPITAL ZENICA

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Introduction: IgG4 – related disease (IgG4-RD) is a condition first described as a form of autoimmune pancreatitis. Today it is known that IgG4-RD has an immune-mediated origin, characterized by a mass-forming lesion, the infiltration of IgG4-positive plasma cells and elevated serum IgG4 levels. Besides pancreatitis, other clinical manifestations include parotid and lacrimal swelling, lymphadenpathy, as well as sclerosing cholangitis and retroperitoneal fibrosis. In this case report we report a symptomatic patient diagnosed with IgG4-RD as a first patients in our hospital with confirmed diagnosis of this auto-immune disease.

Case Report: A 51-year-old female presented herself in emergency ward with abdominal pain dominantly under RCA (right costal arch). She reported that she has had similar symptoms earlier but not that intense. A clinical review is negative for other symptoms. Laboratory findings and ultrasound imaging were interpreted as acute cholecystitis for which she was initially treated conservatively and operated few days after. Few months after that she reported with blurred vision and pain in her left temple. After additional testing, magnetic resonance imaging (MRI) confirmed a tumor-like lesion in her brain. Surgical treatment was performed and confirmed tissue infiltrated with plasma cells. Based on theoretical knowledge, IgG4-RD was considered and plasma levels of IgG4 class were tested highly elevated. Corticosteroid treatment was started with positive response and resolution of symptoms as well as full recovery of the patient.

Keywords: *auto-immune disease, IgG4-related disease, orbital tumor*

[Abstract:1014]

THROMBOTIC THROMBOCYTOPENIC PURPURA

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Classically defined by fever, purpura, thrombocytopenia, anemia, acute kidney injury and neurological symptoms, thrombotic thrombocytopenic purpura (TTP) is a rare and potentially fatal disease if not diagnosed and treated in a timely manner, which is due to loss of of ADAMTS13 synthesis, essential for von Willebrand factor regulation. With congenital or immune etiology, TTP still presents a diagnostic challenge whose early identification is of the utmost importance for the best outcome in the patient's treatment. A 62-year-old, female, presented to the general emergency department with abdominal pain that started 3 days prior, accompanied by a single episode of vomiting, without skin changes. She had severe thrombocytopenia, with a significant increase in total and indirect bilirubin. She tested positive for SARS-CoV-2. The patient was hospitalized for surveillance and study of analytical changes. A peripheral blood smear (PSE) was requested and corticosteroid therapy was instituted. Due to the presence of schistocytes in PSE and the clinic, the hypothesis that it was a TTP was considered, which is why ADAMTS13 research was requested. The patient died during hospitalization, with rapidly progressive clinical worsening, despite the efforts made to reverse the situation. Post-mortem genetic study confirmed ADAMTS13 absent expression. Early diagnosis and immediate treatment are essential to reduce morbidity and mortality in patients with a high clinical suspicion of TTP. The therapeutic approach with plasma replacement technique and surveillance in a specialist consultation for symptomatic surveillance and control of possible triggers that could again lead to the onset of an exacerbation of the underlying disease.

Keywords: *thrombotic thrombocytopenic purpura, plasma exchange therapy, purpura, ADAMTS13*

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[Abstract:1015]

A METHICILLIN SUSCEPTIBLE STAPHYLOCOCCUS AUREUS BACTEREMIA IN A PATIENT WITH METASTATIC CANCER THAT LED TO UNCOVERING OF POSSIBLE IGG4-RELATED DISEASE

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Case Report: We present the case of a 69-year-old patient with retroperitoneal fibrosis uncovered after treatment for MSSA bacteremia.

Description: A 69-year-old male, with a medical history of colon cancer, metastatic to liver and lungs, under chemotherapy, was admitted due to fatigue and fever. Laboratory tests showed elevated inflammatory markers and impaired kidney function. The patient was initially diagnosed with MSSA (Methicillin Susceptible *Staphylococcus aureus*) bacteremia, without endocarditis, and was treated with cloxacillin. However, no significant laboratory or clinical improvement was noted.

Findings: A computed tomography scan revealed the presence of a retroperitoneal mass engulfing the urinary bladder, along with significant renal pelvicalyceal dilatation. A quantitative measurement of serum immunoglobulins was performed, and a high IgG4/IgG ratio (0.35) was calculated. The patient was treated with dexamethasone and consequently had a rapid clinical response, with fever remission, as well as improvement of kidney function and inflammation. A new CT scan revealed considerable reduction of the retroperitoneal mass volume.

Conclusions: Retroperitoneal fibrosis is associated with obstructive uropathy. IgG4 related disease is an immune-mediated fibroinflammatory condition, occasionally associated with malignancies, that accounts for nearly 30 to 60 percent of the total retroperitoneal fibrosis cases and can affect the vessels of the area or the ureters, precipitating obstructive uropathy and renal injury. IgG4 serum values, although not specific, tend to be elevated in two-thirds of these patients. Glucocorticosteroid administration is the first therapeutic choice and response, including reduction of mass, is expected within one to four weeks.

Keywords: *retroperitoneal mass, IgG4 related disease, malignancy, bacteremia, dexamethasone*

[Abstract:1022]

HIP OSTEONECROSIS

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Pain is a common cause of visits to the emergency room, but sometimes they reveal less frequent etiologies.

We report the case of a female patient, 45 years old, with a history of rheumatoid arthritis treated with methotrexate and prednisolone 7.5 mg. She accessed the Emergency Department in August 2022 due to intense pain in the right hip joint, which worsened when the limb was mobilized, causing difficulty in walking, although not disabling. She also reported a fall in January of the sane year, with trauma to the right lower limb and pain beginning at the time of the trauma and continuing ever since, with progressive worsening. Physical examination objectified pain with active and passive mobilization of the right hip joint. A radiograph of the hip joint was performed, which revealed avascular necrosis of the head of the femur. She was discharged with analgesic medication and continued follow-up in consultations.

Keywords: *hip osteonecrosis, corticotherapy, fall*



Figure 1. *Hip radiograph.*

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Figure 2. Hip radiograph.

[Abstract:1030]

UNUSUAL PRESENTATION OF GIANT CELL ARTERITIS

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Case Description: A 58-year-old man with history of hypertension and type 2 diabetes, presented to the Emergency Department with a 2-week developed fever, lower back pain radiating to lower limbs, weight loss and persistent scrotal inflammation not improved with outpatient antibiotic treatment. Laboratory tests revealed a C-reactive protein of 500 mg/dL, erythrocyte sedimentation rate of 110 mm/h, leukocytosis with predominance of polymorphonuclear cells, normocytic anaemia with hemoglobin of 8 g/dL, attributed to the acute inflammatory process, and normal urine sediment. Scrotal ultrasound showed findings consistent with epididymis-orchitis and a treatment with ceftriaxone and doxycycline resolved the symptoms. Five days later, he was readmitted due to recurrent fever and elevated inflammatory markers. Urine and blood cultures and serological tests yielded negative results, prompting evaluation for fever of unknown origin, including PET-CT and temporal artery Doppler ultrasound. PET-CT was normal, while Doppler showed a hypoechoic halo sign suggestive of arteritis. The temporal artery biopsy was compatible with giant cell arteritis. Ophthalmologic examination revealed normal findings. Treatment with intravenous methylprednisolone pulses, followed by oral prednisone was started, resulted in resolution of fever and improvement of laboratory parameters. During the outpatient follow-up, methotrexate was added as corticosteroid-sparing agent. After 3-months of treatment, the patient remains asymptomatic with normal laboratory findings.

Discussion and Learning Points: Giant cell arteritis diagnosis was obscured by the concomitant presence of epididymis-orchitis and the atypical demographic profile of our patient. Furthermore, it is noteworthy that Doppler ultrasound of temporal arteries may be more useful than PET-CT in cases with cranial phenotype.

Keywords: *giant cell arteritis, Doppler ultrasound, hypoechoic halo sign*

[Abstract:1035]

RENAL ACTIVITY IN PET/CT MAY IDENTIFY INSIDIOUS VASCULITIS

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Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis constitutes an important subgroup of small vessel vasculitis. Renal and pulmonary involvement are frequent. In large vessel vasculitis, positron emission tomography (PET)/computed tomography (CT) is used for diagnosis by directly showing increased activity in the vasculature.

Case Report: A 75-year-old male patient with no chronic disease was admitted with anorexia, weight loss, weakness and difficulty in walking for three months, which started after a radiofrequency ablation procedure. Laboratory findings on admission (Table 1) showed severe acute phase response, high sedimentation rate, anemia, renal failure, reversal of albumin to total protein ratio and positive p-ANCA (MPO). Plasma cell count was normal. Bone marrow biopsy was normal and ruled out hematologic etiologies. The PET/CT scan rule out malignancy but revealed intense diffuse metabolic activity in both renal parenchyma and subcentimetric lymph nodes in the hilar and celiac areas. Increased metabolic activity was observed in both hilar and subcarinal non-specific lymph nodes.

Renal biopsy showed crescentic glomerulonephritis. The final diagnosis was microscopic polyangiitis. After medical treatment and plasmapheresis, neuropathic symptoms completely resolved except for local paraesthesia in the right hand. He gained 35 kg over the 40 kg baseline body weight at one year. Laboratory improvement was also achieved.

Conclusions: Vasculitis should be kept in mind when investigating the etiology of weight loss in an elderly patient. In the present example, renal activity in PET/CT showed critical importance by guiding for the renal biopsy and possibly providing early diagnosis, contributing positively to the prognosis of the elderly patient.

Keywords: *anti-neutrophil cytoplasmic antibody (ANCA), positron emission tomography (PET)/computed tomography (CT), vasculitis*

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	Result	Normal range
Leukocytes/mm3	5,400	(3,900-10,800)
Hemoglobin, g/dl	7.1	(13.2-17.7)
Platelets/µL	254,000	(150,000-400,000)
Sedimentation rate, mm/h	115	(0-20)
C-reactive protein, mg/L	137	(0-3)
Urea, mg/dL	43.9	(10-20)
Creatinine, mg/dL	2.93	(0.7-1.2)
Total protein, g/dL	4.8	(6.6-8.7)
Albumin, g/dL	1.5	(3.5-5.2)
Rheumatoid factor	121	(0-14)
Antinuclear antibody	Negative	Negative
P-ANCA (MPO), U/ml	24	<5
Complement 3, g/L	89	(90-180)
Complement 4, g/L	23	(10-40)
Anti-dsDNA	Negative	Negative
24-hour urine protein	324	<450 mg/day
Anti-GBM Ab	Negative	Negative

Table 1. *Laboratory Findings.*
p-ANCA: perinuclear antineutrophil cytoplasmic antibodies, MPO: Myeloperoxidase, GBM: Glomerular basement membrane.

[Abstract:1041]

PODIATRIC STUDY IN PATIENTS WITH RHEUMATOID ARTHRITIS: CLINICAL, ULTRASOUND, AND FUNCTIONAL CHARACTERISTICS OF THE RHEUMATOID FOOT

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The study on podiatric aspects in rheumatoid arthritis (RA) patients aims to comprehend complications affecting the feet, their prevalence, and impact on patients' quality of life to enhance overall management. The objective is to delineate clinical, ultrasound, and functional characteristics of the rheumatoid foot while identifying factors associated with its functional impairment. In this one-month cross-sectional study, 31 RA patients with minimally or non-deforming feet (male-to-female ratio 6.75, mean age 56.52 years) underwent podoscopic examination, including footprint analysis and clinical podiatric assessment. Ultrasound evaluations of the ankle and foot identified synovitis or tenosynovitis. The Foot Function Index (FFI-F) self-questionnaire, Health Assessment Questionnaire (HAQ), and Disease Activity Score (DAS) 28 were employed.
Results revealed pain, ankle pain, and talalgia in 51.6%, 41.9%, and 32.3% of cases, respectively. Varus and valgus deformities were observed in 32.3% and 25.8%. Hallux valgus and Quintus varus were present in 22.6% and 32.3%. Ultrasound detected synovitis mainly at the 2nd metatarsophalangeal joint (35.5%) and midfoot (16.1%). Posterior tibial and fibular tenosynovitis were observed in 9.7% and 12.9%. Appropriate footwear was noted in 29%, and 19.4% received podiatric care.
Conclusions: RA patients commonly report foot pain and deformities, particularly at the ankle and MTP joints, supported by ultrasound findings. Footwear type correlates with ankle pain, while regular podiatric care associates with higher FFI-F scores.

Keywords: *rheumatoid arthritis, podiatric complications, ultrasound assessment, disease activity score, foot function index, synovitis*

[Abstract:1046]

EVALUATION OF THE RELATIONSHIP OF ANTI NUCLEAR ANTIBODY (ANA) AND DENSE FINE SPECKLED STAINING PATTERN (DFS70-LIKE) WITH CLINICAL OUTCOMES IN GENERAL INTERNAL MEDICINE OUTPATIENT CLINIC

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Autoimmune diseases are caused by the development of an immune response against the organism's own tissues and cells and autoantibody tests are of great importance in the diagnosis and follow-up of the treatment of these diseases. Anti-nuclear antibodies (ANA) are important diagnostic criteria in connective tissue diseases. We screened 1500 patients with rheumatologic complaints who presented to the general internal medicine outpatient clinic between 2019 and 2021 for ANA IIF staining pattern. Our aim in this study was to identify patients with Dense fine speckled-DFS70 staining and immunoblot-confirmed anti-DFS70 positive patients and to evaluate the relationship with clinical findings. Of the 1500 patients screened in the study, 111 patients were found to have IIF Dense fine speckled staining pattern (DFS70) and anti-DFS70 antibody was confirmed by immunoblot method in 40 patients. Our study group consisted of 71 patients with ANA IIF Intense fine speckled DFS70 staining pattern, 40 patients with confirmed anti-DFS70 antibody and 30 ANA negative patients in the control group. Of the 40 patients in our study group, 4 patients (28.6%) were diagnosed with fibromyalgia, 3 with probable SLE (21.5%), 2 with urticaria (14.4%), 1 patient (7.1%) with multiple sclerosis, 1 patient (7.1%) with myasthenia gravis, 1 patient (7.1%) with sarcoidosis, 1 patient (7.1%) with Graves' disease, and 1 patient (7.1%) with pulmonary

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thromboembolism. In the literature, the association of Anti-DFS70 positive patients with rheumatologic disease diagnosis is controversial and there are results associated with different systemic inflammatory and allergic diseases.

Keywords: *anti nuclear antibody, autoimmune diseases, anti-DFS*

[Abstract:1047]

UTILITY OF THE RHEUMATOID ARTHRITIS FOOT DISEASE ACTIVITY INDEX (RADAI-F5) IN ASSESSING RHEUMATOID ARTHRITIS ACTIVITY

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Introduction: Rheumatoid arthritis (RA) disease activity assessments often overlook foot symptoms, potentially leaving residual issues in patients deemed in remission. The Rheumatoid Arthritis Foot Disease Activity Index (RADAI-F5) is a recently validated tool designed for specific evaluation of RA activity in the feet. This study investigates the association between RA foot activity and overall RA activity, functional indices, and joint ultrasound findings.

Patients and Methods: Conducted over one month, this cross-sectional study included RA-diagnosed patients. Quality of life was assessed through the Health Assessment Questionnaire (HAQ). RADAI-F5 quantified RA foot activity, and the Foot Function Index (FFI-F) and Fatigue Severity Scale (FSS) assessed foot function and fatigue. Ankle and forefoot ultrasounds were performed.

Results: Of 31 patients (27 females), mean age was 56.52 years ± 12.746 [25-84], with average disease duration of 10.55 years ± 8.481 [1-34]. RA activity was remission (n=10), low (n=3), moderate (n=17), or severe (n=1). Metatarsalgia occurred in 51.6%, talalgia in 13, and ankle pain in 16 patients. Mean RADAI-F5 was 6.226% ± 2.160 [1-9.2]. RADAI-F5 correlated with DAS28vs activity (r=0.8; p=0.0001), fatigue (r=0.3; p=0.05), FFI-F (r=0.5; p=0.001), and synovitis on ultrasound (p=0.03). No association was found with HAQ functional score (p>0.055).

Conclusions: RADAI F-5 correlates with RA activity, synovitis, and tenosynovitis on ultrasound. Its validity in monitoring RA foot activity is confirmed.

Keywords: *rheumatoid arthritis, foot disease, disease activity index, ultrasound, functional assessment, remission*

[Abstract:1048]

COMPARATIVE STUDY OF THE EFFECTIVENESS OF PHARMACOLOGICAL TREATMENTS IN FIBROMYALGIA

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Introduction: Fibromyalgia (FM) is a complex and chronic medical condition characterized by widespread muscle pain, persistent fatigue, and various associated symptoms, including sleep disturbances and cognitive issues. Despite its clinical significance, FM poses challenges in terms of diagnosis and management. Available therapeutic options, such as antiepileptics and antidepressants, primarily aim to alleviate symptoms.

Patients and Methods: This cross-sectional study involved patients diagnosed with FM based on the modified 2016 ACR criteria. The visual analog scale for pain (VAS), the Fibromyalgia Impact Questionnaire (FIQ), and the Fatigue Severity Scale (FSS) were assessed.

Results: Forty-four patients participated, comprising 39 women and 5 men, with a mean age of 56.2±11.37 years. 23.3% of patients had tried pregabalin (n=10), 28 had tried tricyclic antidepressants, and 2 were treated with gabapentin. The median fatigue scale (FSS) was 38.5 [9-62], with 23 patients having an FSS greater than 36, indicating objectively elevated fatigue. Pregabalin use was associated with less intense pain on the VAS (p=0.002). Selective serotonin reuptake inhibitors (SSRIs) were linked to less functional impairment on the FIQ (p=0.011). However, pregabalin was not associated with the measure of functional impact (p=0.121) or fatigue (p=0.177). Gabapentin showed no association with the evaluated variables (p>0.05), and SSRIs had no association with pain (p=0.0837) and fatigue (p=0.603).

Conclusions: Pregabalin benefits FM pain, while SSRIs improve function. Various FM treatments have limited impact on fatigue, remaining an unmet need in FM management.

Keywords: *fibromyalgia, pregabalin, antidepressants, pain management, fatigue severity, treatment efficacy*

[Abstract:1049]

EVALUATION OF ORTHOPEDIC PRACTICES IN PATIENTS WITH CHRONIC INFLAMMATORY RHEUMATISM: RESULTS AND RECOMMENDATIONS

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Introduction: Remarkable progress in the management of chronic inflammatory rheumatism (CIR) has undeniably enhanced

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patients' quality of life. However, for those with advanced joint damage, particularly in conditions like ankylosing spondylitis (AS), rheumatoid arthritis (RA), psoriatic arthritis (PsA), or juvenile idiopathic arthritis (JIA), total hip arthroplasty (THA) and total knee arthroplasty (TKA) remain indispensable in their care.

Patients and Methods: We conducted a self-administered questionnaire distributed to orthopedic surgeons through a form shared on social networks.

Results: Data were collected from 66 orthopedic surgeons, including 3.03% females (n=2) and 96.97% males (n=64). The median age was 32 years [26-45]. The sample comprised 52 residents, 12 assistants, and two associate professors, with a median of 5 years [1-20] of experience. 81.8% performed prostheses on rheumatic joints, with 59.1% acknowledging additional difficulties. Regarding medication management, 71.2% recommended maintaining the usual dosage of disease-modifying antirheumatic drugs (DMARDs) for patients undergoing elective THA or TKA. Biologic therapy recommendations varied, with 53% advocating surgery after the dose cycle. For targeted therapies, 65.2% suggested stopping 3 days pre-operation. All surgeons agreed to resume medications conditionally after the wound showed healing signs, usually around 14 days post-operation. Continuing the current daily dose of corticosteroids (CS) was unanimously recommended.

Conclusions: Prosthetic replacement of rheumatic joints is complex with inherent risks, including infection. The 2022 American College of Rheumatology (ACR) recommendations offer vital guidance, emphasizing the need for rheumatologists to collaborate with orthopedic colleagues for optimal patient management.

Keywords: *chronic inflammatory rheumatism, total hip arthroplasty, total knee arthroplasty, disease-modifying antirheumatic drugs (DMARDs), biologic therapy, American College of Rheumatology (ACR) recommendations*

[Abstract:1064]

A SUSPICIOUS HEPATOCARCINOMA

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Primary biliary cholangitis (PBC) is a rare and chronic inflammatory autoimmune cholestatic liver disease, predominantly affecting women aged 40-60 years. It damages small intrahepatic bile ducts, resulting in fibrosis and, eventually, cirrhosis with associated complications. The clinical manifestations and course vary, with many patients being asymptomatic and diagnosed through a cholestatic biochemical profile.

We report the case of a 62-year-old woman admitted for investigation due to cytocholestasis and a suspicious hepatic nodule, described as a potential hepatocarcinoma diagnosed through abdominal magnetic resonance imaging (MRI) conducted to further evaluate the etiology of cytocholestasis. She was asymptomatic, and no abnormalities were found on physical examination.

Laboratory studies revealed a cytocholestatic biochemical profile. Subsequent abdominal MRI confirmed hepatic cirrhosis without additional findings.

An autoimmune etiology was investigated, revealing antinuclear antibody positivity, along with anti-mitochondrial antibodies, including positive M2 and M2-E3. A liver biopsy was performed, showed chronic hepatitis with moderate density portal infiltrate, predominantly lymphoplasmacytic, non-destructive lymphocytic cholangitis lesions, portal fibrosis with connective septa of variable thickness and nodular outline, marginal necrosis (moderate interface type), moderate-intensity intralobular necroinflammatory lesions.

The patient was diagnosed with PBC and initiated ursodeoxycholic acid, leading to the resolution of the cytocholestatic biochemical profile.

This case highlights the importance of early diagnosis to prevent potential complications and progression to cirrhosis. In this instance, the patient has already been diagnosed at the cirrhosis stage, necessitating regular monitoring. It also underscores the relevance of discussing possible diagnostic hypotheses in the context of cholestasis, ensuring timely and appropriate diagnosis and treatment.

Keywords: *primary biliary cholangitis, cirrhosis, cholestatic liver disease, autoimmune disease*

[Abstract:1070]

PAGET'S DISEASE: EPIDEMIOLOGICAL PROFILE

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Introduction: Paget's Disease, characterized by disordered bone tissue remodelling, is a common osteopathy affecting around 2% of individuals over 50. This study aims to elucidate the epidemiological, clinical, radiological, and therapeutic characteristics of Paget's Disease through an analysis of 81 cases.

Materials and Methods: This retrospective study spans 33 years, involving 81 Paget's Disease cases.

Results: The cohort comprises 54 men and 27 women (age range: 30 to 84, mean age at diagnosis: 54.01 years). The predominant symptom was isolated pain (83.95%), with other presentations including headache with hypoacusis (3.7%), carpal tunnel syndrome (1.23%), meralgia (1 case), bone deformity (2.46%), fracture (1 case), inaugural spinal cord compression (4.93%), and cauda equina syndrome (1 case).

Regarding pain characteristics:

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Mechanical pain occurred in 54 patients.
Mixed pain was observed in 11 patients.
Inflammatory pain was present in 16 patients.
Joint pain affected 47 patients, primarily in the spine (68.08%), hip joint (21.27%), knee (8.51%), and wrist (2.12%).
Radiological exploration identified various manifestations, including ivory vertebrae, cranial vault condensation, and pelvic or femoral osteocondensation. Bone scintigraphy exhibited hyperfixation in multiple locations.
Sixty-nine patients received bisphosphate treatment, with clinical improvement noted in 49 patients, including pain regression and complications amelioration.
Conclusions: Paget's Disease, a benign bone disorder, responds effectively to bisphosphate treatment, guided by evolving bone remodelling markers. Surgical intervention remains a consideration, particularly in cases of joint involvement.

Keywords: Paget's disease, bisphosphate treatment, joint involvement

[Abstract:1075]

FORESTIER'S DISEASE: FOCUS ON EPIDEMIOLOGICAL CHARACTERISTICS AND ASSOCIATED COMORBIDITIES

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Introduction: Forestier's disease, also known as ankylosing vertebral hyperostosis, primarily affects the elderly and those with metabolic vulnerabilities. It remains an insufficiently explored territory, occasionally confused with ankylosing spondylitis and osteoarthritis. The study's objective is to investigate the epidemiological characteristics and explore key risk factors and comorbidities associated with Forestier's disease.
Materials and Methods: Conducted over 18 years at Hedi Chaker University Hospital in Sfax (Tunisia), the study included 63 patients hospitalized for Forestier's disease.
Results: With an average age of 63.36 years, the cohort demonstrated a male predominance (60.31%). Urban and rural origins did not significantly impact disease occurrence. Comorbidities and cardiovascular risk factors included smoking (31.74%), overweight/obesity (28.57%), hypertension (42.85%), diabetes (39.68%), and dyslipidemia (25.39%). Osteoarthritis was prevalent (47.61%), with knee (31.74%), spinal (19.04%), and hip (11.11%) osteoarthritis noted. Gout was observed in 12.69% of cases, and chronic kidney disease in 6.34%. Monitored comorbidities encompassed rheumatoid arthritis, monoclonal gammopathy, and chronic obstructive pulmonary disease.
Conclusions: Forestier's disease manifests predominantly in the elderly, demonstrating a mild male bias. Urban or rural distinctions do not significantly influence its occurrence. Cardiovascular comorbidities, including smoking, obesity, and diabetes, are implicated. Osteoarthritis frequently accompanies Forestier's disease, emphasizing the complex interplay of factors in its genesis.

Keywords: Forestier's disease, cardiovascular risk factors, comorbidities, osteoarthritis association

[Abstract:1082]

THE CLINICAL AND RADIOLOGICAL CHARACTERISTICS OF FORESTIER'S DISEASE

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This study investigates the clinical characteristics of Forestier's disease in a cohort of 63 patients. Patients were admitted to the Rheumatology Department of Hedi Chaker University Hospital in Sfax, Tunisia, over an 18-year period (2004-2022) for Forestier's disease.
In our series, the disease was primarily discovered based on clinical signs in the majority of cases (79.36%, n=50), while 20.64% of cases (13 patients) were asymptomatic, incidentally identified through standard radiographs. The onset of symptoms occurred within one year in 57.14% of patients, between one to two years in eleven patients, and over two years in nine patients.
Back pain was prevalent, affecting 82.53% of cases (n=52), with various spinal levels involved. Mechanical back pain was persistent in 80.95% of cases, paroxysmal in 19.05%, and responsive to first-line analgesics in 95.23% of cases. The pain was mostly mild, with an intensity score below 5 out of 10 in 85.71% (n=54).
Radiculalgia accompanied back pain in 54% of cases (n=27), with sciatica being the most frequent (15.87%). Lumbar spinal stenosis signs were present in 12.69% of patients.
Dysphagia and sleep apnea syndrome were rare but reported in two and one patient(s), respectively. Mechanical arthralgia without arthritis was noted in 15.87% of cases (n=10).
Stiffness was observed in various spinal regions, and two patients exhibited symptoms of spinal cord compression.
Symptomatic forms prevailed in the series, with pain, particularly back pain, being the predominant symptom, while radicular pain was common, and dysphagia was infrequent.

Keywords: Forestier's disease, back pain, radiculalgia, spinal stenosis signs

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[Abstract:1086]

NEPHRITIC SYNDROME IN PATIENT WITH PROGRESSIVE LYMPHOMA AND SJOGREN'S SYNDROME

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Clinical Case: A 54-year-old woman with a history of Sjogren's syndrome and MALT B-cell lymphoma in the submaxillary gland, previously treated with surgery and radiotherapy, presented with recent CT evidence of tumor progression in the right parotid. She arrived at the emergency department with intense discomfort, fatigue, low-grade fever, night sweats, and progressive dyspnea over a month. Examination revealed hypertension, tachycardia, pale mucocutaneous appearance, and bibasilar crackles on respiratory auscultation. Investigations showed left pleural effusion on chest X-ray, and laboratory results indicated renal dysfunction, anemia, leukopenia, and thrombocytopenia. The persistence of hypertension prompted an internal medicine consultation. Peripheral blood smear revealed schistocytes, leading to a diagnosis of thrombotic microangiopathy (TMA), ruling out thrombotic thrombocytopenic purpura (normal ADAMTS13 activity). Skin lesions suggestive of cutaneous vasculitis and echocardiography indicating a dilated left ventricle with slightly reduced ejection fraction were observed.

Hypothesis: The causes considered in the differential diagnosis of our patient's TMA are complement-mediated TMA and secondary causes related to connective tissue disorders, neoplasia, or malignant hypertension.

Diagnostic Pathways: Ophthalmic examination rules out malignant hypertension. PET-CT indicates disappearance of cervical lymphadenopathy. Anti-complement antibodies and genetic test complement pathway testing are negative. Subsequent renal biopsy reveals membranoproliferative glomerulonephritis with immune deposits, suggesting the possibility of cryoglobulins, which had previously tested negative.

Immunological parameters and histology support an autoimmune etiology, leading to treatment with steroids, plasma exchange, human immunoglobulins and rituximab.

Discussion and Learning Points: This case underscores the diverse presentations of systemic diseases, necessitating a multidisciplinary approach for accurate diagnosis and effective treatment.

Keywords: *thrombotic microangiopathy, Sjogren's syndrome, MALT B-cell lymphoma, nefritic syndrome, cryoglobulins*

[Abstract:1089]

THE INTEREST OF SELF-ASSESSMENT SCALES IN THE FOLLOW-UP OF SYSTEMIC SCLERODERMA PATIENTS

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Introduction: Scleroderma is a connective tissue disease that primarily affects adult women. It is a systemic disease leading to severe and potentially irreversible damage that impairs patients' quality of life.

Methods: This is a cross-sectional study conducted during July 2023 in our department. We interviewed patients using two validated scales: the Cochin functional hand scale and the scleroderma HAQ. Patient data were retrieved from charts.

Results: 25 patients were assessed aged between 38 and 76 years. Among our patients, 10 had diffuse skin involvement, and 9 had a history of ischemic digital ulcerations 20 patients had pulmonary involvement. The Cochin functional hand scale covers 5 areas of life and contains 18 items. The overall score reflects the degree of functional impairment. The most impaired areas were dressing and office activities. The presence of digital ulcers or scarring was associated with impairment of delicate tasks. Finally, the area least impaired was toileting. The scleroderma HAQ is a self-assessment score for the initial assessment and follow-up of patients. The tasks with the greatest level of difficulty were gardening, followed by climbing stairs. 12 of our patients needed help with walking and shopping, 7 patients needed help with hygiene.

The pain caused by the disease was severe or very severe in 19 patients, related to pulmonary involvement in 13 patients, bowel involvement in 6 and to digital ulcerations in 4.

Conclusions: A comprehensive assessment that takes into account the patient's experience is necessary to evaluate the quality of life and the impact of this disabling disease.

Keywords: *scleroderma, disability, scores*

[Abstract:1105]

ACUTE CENTRAL NERVOUS SYSTEM EVENT IN THE CONTEXT OF HYPOCOMPLEMENTEMIA URTICARIAL VASCULITIS?

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Introduction: Urticarial vasculitis includes clinical manifestations of urticaria and histopathologic evidence of cutaneous leukocytoclastic vasculitis of the small vessels. It could affect only the skin or other organ systems.

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Clinical Case: A 46 years old Brazilian woman presented with a previous history of hypertension diagnosed 10 years earlier and urticaria associated to angioedema with suspected hypocomplementemia urticarial vasculitis under study. She went to the Emergency Department due to headache different from usual. On objective examination, the patient was hypertensive (219/130 mmHg), tachycardic (123/min) and subfebrile. She had urticarial lesions on her lower limbs. Analytically with slight leukocytosis (13800). Urine examination showed sub nephrotic proteinuria (3.1g/dL) without hematuria. Electrocardiogram showed no evidence of acute ischemia. Brain computed tomography showed nonspecific hypodensity in the left striatal region. From tests carried out during hospitalization, the patient had positive ANA, complement deficiency and decreased C1q. Positive Schimer test. Biopsy of skin lesion with cutaneous small vessel vasculitis/leukocytoclastic vasculitis. As a complication, the patient presented a difficult-to-control hypertensive emergency with associated with dysarthria and motor deficit in the left lower limb. Imaging with new acute intracranial lesions. There was no improvement with corticosteroid therapy, but plasmapheresis was performed, and the condition improved. She remained weaned from corticosteroid therapy and monitored in consultation.
Conclusions: Hypocomplementary urticarial vasculitis can be systemic. However, there is no evidence of acute CNS involvement with stroke. In this case, we had a hypertensive context that may be associated with vasculitis and have led, as a consequence, to stroke.

Keywords: *hypertensive emergency, urticarial vasculitis, acute central nervous system event*

[Abstract:1109]

THE CLINICAL DIAGNOSIS OF POLYMYALGIA RHEUMATICA AND GIANT CELL ARTHRITIS IN THE EMERGENCY DEPARTMENT

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Introduction: Polymyalgia rheumatica is an inflammatory disease that mainly affects women over 50 years of age. The diagnosis is clinical, based on the involvement of the shoulder and pelvic girdles with morning stiffness and associated constitutional symptoms that respond favorably to corticosteroid therapy. In 50% of cases, it may be associated with giant cell arthritis.
Clinical Case: An 85 years old female with previous history of hypertension, dyslipidemia and chronic bronchitis. Medicated with antihypertensive drugs, statins and bronchodilators. In a previous analytical study, the patient presented elevated erythrocyte sedimentation rate (120 mm), normocytic normochromic anemia and inflammatory hyperferritinemia. She went to the Emergency Department for episodes of syncope. Concurrently with 1 to 2 months of evolution of intense right frontotemporal headache with sudden ipsilateral vision loss, pain of inflammatory characteristics and morning stiffness, associated with significantly weight loss, anorexia, nausea and vomiting. Physical examination reported no relevant alterations.
A biopsy of the right temporal artery was scheduled, and high-dose corticosteroid therapy (prednisolone 50 mg per day) was initiated. Temporal artery biopsy showed histomorphological changes characteristic of giant cell arteritis. Angiography showed thickening of the thoracic aorta. Patient with significant clinical improvement after treatment.
Conclusions: This clinical case demonstrates that sometimes the reasons that lead patients to the Emergency Department hide a less evident but more relevant clinical context. Thus, it is important to take an appropriate history with a high level of suspicion for a proper diagnosis and treatment.

Keywords: *polymyalgia rheumatica, giant cell arthritis, headache*

[Abstract:1215]

A RARE CASE OF INFLAMMATORY MYOPATHY IN A PSYCHIATRIC PATIENT

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Inflammatory myopathies are rare autoimmune diseases that involve chronic muscle inflammation and are associated with motor weakness of varying severity.
72-year-old woman, with history of Diogenes' syndrome, arterial hypertension and dyslipidaemia medicated with atorvastatin. The patient presented multiple times to the emergency department with myalgias and weakness in the lower limbs causing frequent falls and progressive loss of autonomy. Blood tests showed elevated creatinine kinase 800 U/L (CK) and the statin was suspended. Two months after the first admission, she was hospitalized due to fluctuations in consciousness and paraparesis. Initial blood analysis showed CK 3610 U/L, myoglobin 6271 ng/mL, reactive protein C 290 mg/dL, aspartate aminotransferase 862 U/L, alanine aminotransferase 339 U/L, lactate dehydrogenase 3820 U/L. Progression of tetraparesis with associated hyperreflexia and dysphagia was observed. The etiological study included: muscle magnetic resonance with marked myositis involving the gluteal muscles, the infraspinatus and supraspinatus and the thighs; electromyography showing myopathy and signs of muscle fibre necrosis predominantly on proximal muscles; elevated angiotensin-converting enzyme; lumbar puncture, autoimmune study including anti-3-hydroxy-3-methyl-glutaryl-coenzyme A reductase (anti-HMGCR), protein electrophoresis, and thoraco-abdomin-pelvic computerized tomography were normal. The hypothesis of inflammatory myopathy/myositis was put forward and a muscle biopsy was performed which revealed fibre atrophy and necrosis, suggesting an immune-mediated necrotizing

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myositis. Corticoids were initiated with resolution of dysphagia and slight improvement of tetraparesis, and the patient was referred to neuromuscular disease consultation.
In this case there was a delay in diagnosis because the patient's symptoms were not evaluated due to her psychiatric history, showing the importance of a thorough clinical history and etiological investigation.

Keywords: *inflammatory myopathy, psychiatric disease, Diogenes' syndrome*

[Abstract:1225]

THE ENIGMA OF FEVER AND VIOLET SKIN LESIONS

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A 33-year-old woman with a history of subacute thyroiditis and right supraorbital granuloma secondary to sarcoidosis presented to Internal Medicine with symptoms initiated by a hiking trip in February. She complained of hyperalgesia in the first phalanx of the left foot radiating to the metatarsals, along with hypoesthesia and erythema on the heel. Additionally, violaceous papules appeared on the left foot, left leg, and right hand. Experiencing exertional dyspnea, nocturnal fever, and diminished breath sounds at the left lung base, she was admitted.
Physical examination revealed violaceous lesions on the left foot, shin, and knuckles of both hands. Initial bloodwork showed leukocytosis with eosinophilia. Serological tests for atypical pneumonia, Rickettsia conorii, and Coxiella burnetii were negative. Autoimmunity parameters revealed a positive c-ANCA pattern.
Chest X-ray indicated bilateral pleural effusion and mediastinal widening, prompting a chest CT that revealed lymphadenopathy, bronchiectasis, and pleural effusion. An echocardiogram detected pericardial effusion, leading to EBUS for biopsies to rule out sarcoidosis.
Skin biopsy confirmed granulomatosis with polyangiitis (GPA). Treatment with corticosteroids and immunoglobulins showed a favorable response.
GPA, a medium-sized vessel vasculitis, presents with diverse symptoms. Diagnostic criteria include eosinophilia, asthma history, sinus pain, mononeuropathy, and migratory lung infiltrates. Treatment involves corticosteroids and additional immunosuppressants if needed. Recognizing the complexity of GPA is crucial for accurate diagnosis and prompt intervention.

Keywords: *c-ANCA, vasculitis, eosinophilia*



Figure 1.

[Abstract:1236]

EOSINOPHILIA: FROM ASTHMA TO SMALL VESSEL INVOLVEMENT. KEYWORDS: ASTHMA, EOSINOPHILS, ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)-ASSOCIATED VASCULITIS; EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA); LUNG INFILTRATES

Patricia Urrutia López, Laura León Ruiz, Ana Galdeano Escanez, Marta Segura Diaz, Luisa Flores Serrano

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A 53-year-old male with a history of chronic rhinitis/sinusitis, polyps, and severe eosinophilic asthma. He was admitted due to painful distal paresthesias, general discomfort, and fever. Physical examination revealed distal weakness, sensory alteration, bilateral wheezing on lung auscultation, and palpable purpuric lesions on forearms. Complementary tests showed axonal sensory-motor mononeuritis, eosinophilia, VSG 60 mmH, troponin 688, proBNP 159; Immunophenotype: no lymphoid cells suggestive of hypereosinophilic syndrome; ANCA 1/160 (MPO 162), chest CT with bilateral patchy infiltrates, and nasal mucosa biopsy vasculitis and eosinophilic granulomatosis. He was diagnosed with severe eosinophilic granulomatosis with polyangiitis (EGPA).
He was initially treated with glucocorticoids and cyclophosphamide, with a partial response, so was changed to rituximab and mepolizumab.
EGPA is a rare multisystem disorder characterized by the triad of severe asthma, peripheral eosinophilia, and small to medium vessel vasculitis. It typically progresses through an initial prodromal/allergic phase, followed by an eosinophilic and finally a vasculitic phase. It presents around the age of 40 with no gender predominance. Its pathogenesis is unknown. 30-60% have ANCA+ against myeloperoxidase; in these patients, purpura, mononeuropathy, and alveolar hemorrhage are more common, while ANCA- patients often exhibit eosinophilic manifestations such as cardiac.
The most common manifestations are respiratory (90%), followed

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by neuropathies cutaneous, and cardiovascular. Diagnosis is based on clinical criteria, eosinophilia and histological findings. Cardiac involvement is the most serious. Therefore, as part of disease evaluation, active monitoring of cardiac involvement is crucial. The mainstay of treatment is glucocorticoids combined with immunosuppressants or biologics, with mepolizumab (anti-IL-5 monoclonal antibody).

Keywords: *asthma, eosinophils, anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, eosinophilic granulomatosis with polyangiitis (EGPA), lung infiltrates*



Figure 1. *Chest x-ray.*

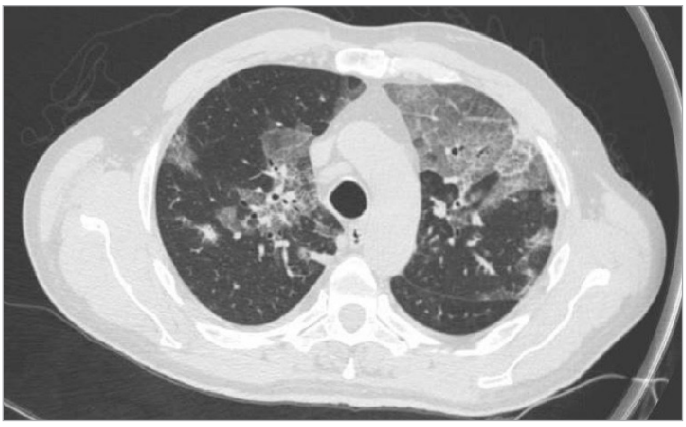


Figure 2. *CT scan.*



Figure 3. *Purpuric lesions.*

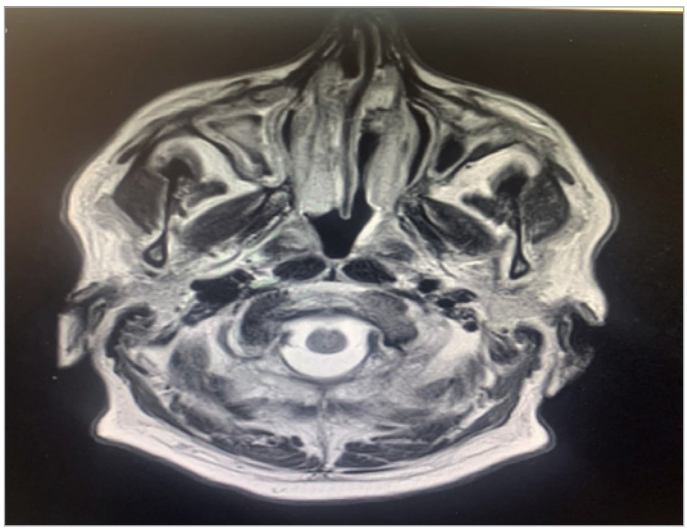


Figure 4. *RMN.*

[Abstract:1244]

A PECULIAR HEADACHE

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A 67-year-old man, without medical history, was admitted to the Internal Medicine service due to a holocranial headache for the last month, along with evening fever, jaw claudication and a weight loss of 10 kg. No loss of vision.

At the examination level we observed the presence of pulsating, beaded temporal arteries.

In the blood test an increase in inflammatory parameters, ESR 84.

The first imaging test performed was a bilateral temporal artery Doppler, with changes consistent with temporal artery arteritis.

CT of thorax and abdomen, with discovery of a focal lesion in the neck of the pancreas.

Given these findings, PET-CT was performed and a slight increase in metabolism was observed in the pancreatic head and increased metabolism in the left temporal artery, and both vertebral arteries compatible with vasculitic activity.

Finally echoendoscopy was performed with a diagnosis of a moderately differentiated invasive ductal adenocarcinoma of the pancreas.

We obtained a diagnosis of giant cell arteritis and concomitant pancreatic neoplasia.

Treatment with corticosteroids was started immediately, leaving the patient completely asymptomatic. Regarding of pancreatic neoplasia said, it was presented to the oncological committee, and it was decided to perform Whipple surgery and adjuvant chemotherapy.

Currently the patient is asymptomatic, with oncology follow-up.

According to the scientific evidence, we can conclude that concurrent malignancy in temporal arteritis is not a rare finding, being observed in up to 7.4% of the cases described in the literature. Patients with and without malignancy appear almost

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indistinguishable in the characteristics and outcome of vasculitic disease.

Keywords: *headache, arteritis, neoplasia*

[Abstract:1247]

MALE WITH SYMMETRICAL POLYARTHRALGIAS AND DYSPNEA

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We present the case of a 65-year-old man, smoking 1 pack/day.
He consulted to Internal Medicine due to symmetrical migratory polyarthralgias (large and small joints), asthenia, dyspnea with exertion and dry cough, without fever of 2 months' duration.
He presented edema in the metacarpophalangeal joints and pain with active and passive joint mobilization.
Laboratory tests: leukocytosis, neutrophilia, CRP 215 mg/dL, anemia and thrombocytosis. The examination was expanded to rule out infectious etiology, without microbiological isolation. A chest x-ray revealed a nodular image in the right lower lobe and patchy infiltrates in the upper lobes. We were impressed by an autoimmune etiology with low suspicion of an infectious etiology.
The autoimmune study reveals elevated ESR (64 mm/h), as well as RF and anti-CCP at high titers, with negative ANA, all of which is suggestive of rheumatoid arthritis (RA). We started prednisone 30 mg/24h and NSAIDs for symptomatic control, with slight improvement initially. However, subsequent worsening occurred, so methylprednisolone was started in boluses of 125 mg IV for 3 days.
Given the symptoms of dyspnea and dry cough, we performed a chest CT (two subcentimeter nodules in middle lobe, consolidations in upper left lobe and subpleurals) compatible with pneumonia. However, the patient remained afebrile, without expectoration or other findings that support infectious pneumonia. Correlating the symptoms, the most likely diagnosis was organizing pneumonia. Bronchoscopy was performed with BAL without microbiological isolation, which supported the diagnosis of diffuse interstitial lung disease (DILD) in this context.
Finally, he received methotrexate with spectacular improvement.

Keywords: *rheumatoid arthritis, diffuse interstitial lung disease, organizing pneumonia*

[Abstract:1251]

MULTIDISCIPLINARY UVEITIS UNIT: DESCRIPTIVE STUDY

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Aim: Describe the clinical, etiological and therapeutic characteristics of the patients monitoring in the multidisciplinary uveitis unit of our hospital. Describe the role of Internal Medicine Service in the diagnosis, treatment and control of drug toxicity.
Materials and Methods: Observational, descriptive study, with a review of a series of cases diagnosed of uveitis monitoring in the multidisciplinary consultation of Ophthalmology-Internal Medicine between 2019-2021.
Results: Of the 64 patients treated in the Ophthalmology-Internal Medicine multidisciplinary unit, 50 were diagnosed of uveitis and included in the study. The mean age of onset was 40 years, mostly women (66%). The most frequent forms were anterior location, unilaterality and recurrence. Recurrent forms are more frequent among those with an immune-mediated mechanism. Regarding the etiology, 28% were immune-mediated, 16% infectious, and 34% unclassified. 8% of patients received a different initial diagnosis than the final. The response to treatment has been complete in more than 70%. The main prescription was topical corticosteroids (>90%). Only 2% required surgery, corresponding to unclassifiable etiology. Internal Medicine Service contributed in 84% to establishing the etiological diagnosis and in more than 50% of the patients participated in the control of treatment toxicity.
Discussion: Both the location of uveitis and the establishment of a correct etiology are of vital importance, as they will help in choosing an appropriate treatment. Therefore, multidisciplinary uveitis consultations allow homogenization of the management of patients with this pathology, as well as an early diagnosis and more complete and safe therapeutic management.

Keywords: *uveitis, multidisciplinary uveitis unit, immune-mediated*

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[Abstract:1261]

EVALUATION OF THE RELATIONSHIP BETWEEN DIET, DISEASE SEVERITY AND COLCHICINE RESISTANCE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Aim: Familial Mediterranean Fever (FMF) is a genetic and autoinflammatory disease associated with chronic inflammation. Colchicine is the basis of treatment. The phenotype and severity of FMF are mainly determined by MEFV genotypes, but attacks can be triggered by factors such as diet, intestinal microbiota, stress. We aimed to investigate the relationship between diet and disease severity and colchicine resistance in FMF patients.

Materials and Methods: The study included 153 FMF patients and 105 healthy control groups. FMF patients were grouped according to the presence of colchicine resistance and the severity of the disease was determined by the ISSF score. The frequency of food consumption and the Mediterranean diet adherence questionnaire (KIDMED) were administered to both patients and healthy individuals. The dietary inflammatory index (DII) and dietary acid load (PRAL, NEAP) were calculated and compared.

Results: The KIDMED value was found to be significantly higher in FMF patients compared to the control group. No significant differences were observed among groups in terms of DII, PRAL and NEAP values. It was observed that the urinary pH value was significantly lower in patients compared to the control group and in those with colchicine resistance compared to those without. No relationship was detected between DII, PRAL, NEAP results, and urinary pH.

Conclusions: The severity of FMF may be affected by environmental factors along with genetic dominance. Diet may contribute to inflammation and disease severity in FMF patients.

Keywords: colchicine resistance, dietary, FMF

	Patient (n=153)	Control (n=105)	p
Age, Avg±SS	31.99±10.88	29.56±7.10	0.588*
Height (cm), Avg±SS	167.37±9.52	168.97±9.49	0.253**
Weight (kg), Avg±SS	69.33±15.73	69.82±15.98	0.970**
BMI (kg/m2), Avg±SS	24.65±4.63	24.24±3.88	0.442**

Table 1. *Comparison of demographic data and anthropometric measurements of the groups.*

	Patient (n=153)	Control (n=105)	p*
Creatinine, Avg±SS	1.33±5.97	0.75±0.14	0.961
AST(u/l), Avg±SS	17.50±10.28	16.69±7.89	0.829
ALT(u/l), Avg±SS	29.16±25.85	26.72±17.46	0.420
WBC(x109/l), Avg±SS	7.06±2.07	7.07±1.92	0.732
HGB(g/dl), Avg ±SS	13.55±1.82	14.15±1.70	0.008**
ESH(mm/h), Avg±SS	12.67±9.35	7.69±4.87	0.02
CRP(mg/l), Avg±SS	12.35±28.83	2.87±5.96	<0,001
SAA(mg/dl), Avg±SS	5.25±11.46	-	-
Fibrinogen(g/l), Avg±SS	3.25±0.85	-	-
Urinary pH, Avg±SS	5.90±0.72	6.20±0.86	0.024

Table 2. *Comparison of laboratory values between groups.*

	Patient (n=153)	Control (n=105)	p*
DII, Avg±SS	4.53±0.90	4.64±1.11	0.404
PRAL, Avg±SS	-4.57±24.49	-4.53±21.96	0.942**
NEAP, Avg±SS	42.41±15.56	41.51±13.07	0.627
KIDMED, Avg±SS	5.98±2.17	4.58±2.34	<0.001

Table 3. *Comparison of DII, PRAL, NEAP and KIDMED values of the groups.*

[Abstract:1269]

ERYTHEMA NODOSUM WITH ATYPICAL PRESENTATION

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We present the case of a 68-year-old woman with no drug allergies, a history of hypertension, dyslipidemia and multinodular goiter. She had recently taken a trip to Turkey, in an urban area, with the onset 2-3 weeks after the trip of a picture of nodular, erythematous and painful lesions, initially on the upper extremities and later on the lower extremities. After a few days they changed color, darkening, and pain decreased. The condition was associated with asthenia and arthralgias that were relieved with compression stockings and analgesia, not accompanied by fever. One week before the onset of the lesions she presented self-limited diarrhea, without weight loss.

Examination revealed the previously described lesions on the anterior aspect of the legs and arms, with involvement of the hands and palmar exanthema with bilateral desquamation.

Atypical erythema nodosum, Sweet's syndrome and infectious panniculitis were suggested as differential diagnosis.

Laboratory tests were requested with normal blood count, biochemistry, autoimmunity, TSH, and urine system. Fecal calprotectin 227.3 mg/kg.

Negative serologies for hepatitis, syphilis, HIV, mycoplasma, Chlamydia, Coxiella, Epstein-Barr virus, Borrelia and Brucella. Interferon-gamma was negative.

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Chest X-ray was normal. Colonoscopy was performed, with normal result, which ruled out associated intestinal disease.
A consultation with dermatologist was performed and a skin biopsy confirmed the diagnosis of atypical erythema nodosum. Treatment was with topical corticosteroids, with improvement of the lesions.

Keywords: *erythema, nodosum, atypical, nodular*

[Abstract:1275]

ISCHEMIC STROKE OF UNUSUAL CAUSE

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A 31-year-old black male with no cardiovascular risk factors presented to the emergency room with a sudden onset of pain in the back of the neck, dizziness, nausea, and vomiting. Neurological examination suggested centrally located vertigo, and vertebrobasilar stroke was suspected, but a CT scan and angiogram revealed no findings.
Blood tests, including autoimmunity and serologies, and a normal echocardiogram were conducted. An MRI confirmed an impairment in the restriction at the level of the left posteroinferior cerebellar artery. Neurosonographic study showed an increase in intima-media thickness in the left carotid bulb, possibly related to dysplasia, and asymmetry in the vertebral arteries consistent with left vertebral artery dissection. An angiogram of the renal arteries was requested and showed pathological findings.
Fibromuscular dysplasia is an idiopathic, non-inflammatory, and non-atherosclerotic segmental disorder affecting small and medium-sized arteries. The proliferation of smooth muscle and connective tissue leads to architectural abnormalities in the vessels. It is more common in renal arteries, followed by extracranial cerebrovascular arteries.
The condition predominantly affects young women with an average age of 52, but it can occur at any age. In men, especially smokers, the disease may present more severely.
Diagnosis is now typically made through imaging, such as CT or MRI, showing the characteristic "string of beads" appearance in the multifocal type, or through invasive procedures when intervention is considered.
Treatment depends on the clinical presentation and may involve either conservative or invasive approaches.

Keywords: *cerebral stroke, displasia, central vertigo, young patient, secondary hypertension*



Figure 1. *Renal dysplasia.*

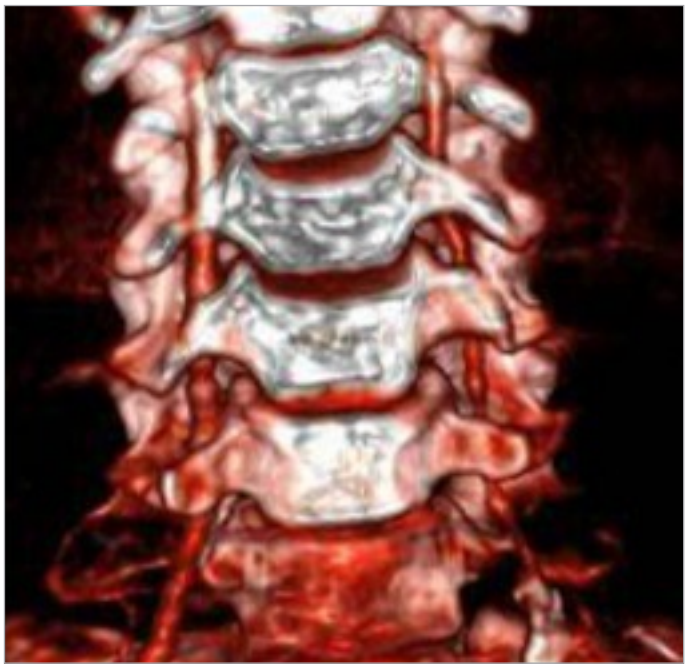


Figure 2. *Vertebral artery dysplasia.*

[Abstract:1279]

MYASTHENIA GRAVIS WITH ATYPICAL PRESENTATION

Genma De Dios Cancelo, Ana Nieto De Pablos, Marina Cazorla González, Paula Celis Sánchez, Tamara Gómez Traveso

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An 87-year-old woman, with a personal history of arterial hypertension and gastroesophageal reflux disease, consulted for a 6-month history of progressive dysphagia with weight loss of 5 kg. The patient reported an ascending oppressive sensation at the

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epigastric level, without chest pain, which did not subside with omeprazole or antiacids. On examination there were no findings of interest, except for slight right palpebral ptosis. The differential diagnosis was esophageal tumor, atypical tumor, neurological disorder and autoimmune disorder. Laboratory tests were performed, with normal blood count, biochemistry, lipid and iron profile, proteinogram, TSH and PTH. Urine and fecal occult blood tests were negative. The test for helicobacter pylori was negative. Hepatitis B, hepatitis C, HIV, syphilis and *Borrelia burgdorferi* serologies were also negative. Interferon gamma was negative. CT was performed without findings of interest and gastroscopy with incipient esophageal ring and normal esophageal, gastric and duodenal mucosa, which did not justify the clinical picture, initially ruling out a tumor. The electrocardiogram was also normal. The patient was re-interviewed and reported progressive asthenia since the onset of the symptoms, slurred voice, difficulty in chewing, and difficulty in eye opening that did not increase with blinking. Symptomatology did not worsen throughout the day and did not improve with rest. The symptoms were not accompanied by dyspnea or claudication of any limb. An autoimmunity study was performed, obtaining positive anti-acetylcholine receptor antibodies, confirming the diagnosis of myasthenia gravis with bulbar involvement. She was treated with pyridostigmine, with favorable evolution.

Keywords: *myasthenia, dysphagia, bulbar*

[Abstract:1298]

ABOUT A CASE: INTRA-ALVEOLAR BLEEDING DURING SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction: Interalveolar bleeding (HIA) is a rare manifestation of systemic lupus erythematosus; a diagnostic and therapeutic emergency with high mortality even with well-adapted treatment.
Case Report: We report the case of a 24-year-old woman, known for systemic lupus erythematosus for 2 years.
Results: The patient was hospitalized for chest pain and low-abundance hemoptysis as part of a deterioration in the general condition. The clinical examination found chest pain mainly on the right; dyspnea to effort; hemopoic spat with auscultation of bilateral crackling rays without fever.
The biology found an aggravation of normochrome normocytic anemia (drop of 03 g/dl) with a negative coombs test; inflammatory syndrome; C3 collapsed, positive FANS and a negative SAPL balance. On chest X-ray: basal alveolar opacity bilateral. The echocardiogram is without abnormalities. Broncho-alveolar washing found siderophages.
We took the diagnosis of HIA on a clinical argument spindle; biological and radiological. The patient benefited from taking immediately three bolus corticosteroid (15 mg/kg/d) then relay per 01 mg/kg/d of prednisolone equivalent and cyclophosphamide bolus (Endoxan®) 500 mg each 15 days for three months; a good development was noted.
Conclusions: HIA is a rare and serious event during LES, a diagnostic and therapeutic procedure must be done urgently to prevent the risk of unfortunate prognosis and high mortality.

Keywords: *intra-alveolar bleeding, systemic lupus erythematosus, broncho-alveolar washing*

[Abstract:1318]

ANTI-IL1 MOLECULE FOR IDIOPATHIC RECURRENT PERICARDITIS

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Case Presentation: The patient was a 40-year-old woman, allergic to metamizole, with no known cardiovascular risk factors. She had suffered from acute recurrent pericarditis in 2015. At diagnosis, the patient reported central thoracic discomfort with ECG changes, and laboratory tests showed elevated CRP and ESR. At discharge, anti-inflammatory drugs and colchicine were prescribed. In a subsequent admission (> 4 weeks without symptoms), she presented an episode of cardiac tamponade and was admitted to the ICU and pericardial drainage was performed. Fluid analysis, autoimmunity and serology were negative but quantiferon was positive, so treatment was started with quadruple therapy with little improvement. After several relapses with corticosteroids and colchicine, treatment was started with boluses of methylprednisolone, azathioprine, colchicine and hydroxychloroquine with good response but relapsed after lowering corticosteroids. In 2021 she started treatment with IL-1 receptor antagonist with good clinical response and no recurrences.
Clinical Judgment: Idiopathic recurrent pericarditis.
Discussion: recurrent pericarditis is defined as a new episode of pericarditis after 4-6 weeks without symptoms. Our patient failed first line, second line and even some third line treatments such as azathioprine. The objective of the treatment of any acute pericarditis is the prevention of recurrences which occurs in 30% of cases. Even though colchicine prevents them in many cases, patients require corticosteroids, which are not free of long-term second effects. The efficacy of anakinra is supported by the AIRTRIP study (2016) and the results of the IRAP registry published in 2020, in which anakinra is effective and safe.

Keywords: *pericarditis, relapses, interleukin-1 receptor antagonist*

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[Abstract:1335]

PROGNOSTIC FACTORS FOR GIANT CELL ARTERITIS

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Introduction: Giant cell arteritis (GCA) is a vasculitis that can have serious functional and vital consequences. Their frequencies are not well known in Tunisia. This study aims to identify the frequencies of complications in our patients.

Methods: We conducted a retrospective analysis of the files of patients with GCA, hospitalized between January 2000 and June 2023. We noted the evolving data in these patients.

Results: We collected 141 patients whose average age was 71.71 ± 8.41 years. There were 68 women and 73 men (F/M sex ratio = 0.93). Joint damage, leading to loss of autonomy, was present in 42 patients (29.8%). Ischemic ocular damage compromising vision was present in 67 patients (47.5%). Aortitis was observed in one case. One patient presented with an abdominal aortic aneurysm concomitant with the positive diagnosis of his vasculitis and without hemorrhagic complications. Three patients had a history of ischemic stroke that preceded the diagnosis of GCA. These complications occurred before the start of treatment in 138 cases (97.87%) and during treatment in 3 cases (2.13%). This treatment was based on corticosteroids in 141 cases (100%) and immunosuppressants in 40 cases (28.36%). The evolution of these complications was towards improvement in 117 cases (82.29%), stabilization in 21 cases (14.89%) and worsening in 3 cases (2.13%).

Conclusions: Although rare, the complications of GCA are serious. They should be systematically searched for in order to improve the prognosis.

Keywords: *giant cell arteritis, vasculitis, horton*

[Abstract:1340]

CRYOGLOBULINEMIC VASCULITIS AND ANCA VASCULITIS: A REAL ASSOCIATION?

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Introduction: The coexistence of granulomatosis with polyangiitis (GPA) and cryoglobulinemic vasculitis is rare and presents a diagnostic and therapeutic challenge.

We report the case of a patient with both cryoglobulinemic vasculitis and GPA.

Observation: This was a 53-year-old patient with no notable history who consulted for vascular purpura of the lower limbs. She reported paresthesias of the lower limbs for two months with episodes of recurrent nasal obstruction and epistaxis. Physical examination revealed a febrile, asthenic patient with vascular-like purpura of both lower limbs with skin ulcerations. Biology showed a biological inflammatory syndrome. The electromyogram of the lower limbs noted a length-dependent axono-demyelinating sensory-motor polyneuropathy. The thoracoabdominopelvic CT scan revealed excavated pulmonary nodules. The skin biopsy showed an appearance of pyoderma gangrenosum. The search for cryoglobulinemia was positive of the IgG, IgA type. The diagnosis of cryoglobulinemia vasculitis was made. The patient was treated with corticosteroid therapy of 60 mg per day and courses of cyclophophamides of 0.7g/m²/SC were initiated. Given the presence of mediastinal adenomegaly, a biopsy revealed giant cell granulomas. Bronchoalveolar washing showed a Gold score of 150 confirming intra-alveolar hemorrhage. The facial CT scan showed septal deviation. The nasal biopsy showed leukocytoclastic vasculitis. The bone marrow biopsy has no tumor cells. The immunological assessment showed the positivity of C-ANCA with anti PR3 specificity. Faced with these clinical and para-clinical arguments, the GPA diagnoses associated with cryoglobulinemia were retained. The cyclophosphamide courses were continued according to the GFEV scheme for vasculitis. The clinical and radiological evolution was favorable.

Keywords: *cryoglobulinemia, GPA, ANCA*

[Abstract:1341]

COMPARISON OF THE CHARACTERISTICS OF GIANT CELL ARTERITIS ACCORDING TO AGE AND GENDER

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Introduction: Giant cell arteritis (GCA) is a predominant vasculitis in elderly patients. This study aims to analyze the characteristics of GCA according to age and gender, highlighting clinical differences between subgroups.

Methods: We conducted a retrospective descriptive study covering the files of patients with GCA, over a period from January 2000 to June 2023. Patients were divided into subgroups according to their age and gender.

Results: A total of 141 was collected. The average age of patients was 71.71 ± 8.41 years, with a balanced distribution between genders (51.8% men and 48.2% women). The average age of discovery in men was 72.09 ± 8.54 years comparable to 71.30 ± 8.30 years in women. Joint involvement was less common in men (n=18) compared to women (n=24). Ocular involvement was more common in men (n=37) compared to women (n=30). Liver damage such as anicteric cholestasis was less common in men (n=4) compared to women (n=7).

Conclusions: This study highlights the importance of considering

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age and gender in the assessment of GCA. The results suggest that clinical features vary depending on these factors, which could have implications for the diagnosis and treatment of this disease.

Keywords: *giant cell arteritis, vasculitis, gender, age*

[Abstract:1346]

AGOSTAR PROTOCOL: EFFECTS OF A STANDARDIZED ACUPUNCTURE PROTOCOL ON CHRONIC PAIN, INFLAMMATION AND LIFE QUALITY IN SYSTEMIC AUTOIMMUNE DISEASES

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We aim to evaluate the efficacy of a standardized acupuncture protocol in a cohort of patients with autoimmune diseases on chronic pain, inflammation and quality of life.

We enrolled 19 female patients from the Policlinico Hospital (Milan), with an established diagnosis of a systemic autoimmune disease and chronic pain with VAS ≥ 3. Patients were treated with a total of 12 acupuncture sessions, during a period of seven months, with a standardized procedure.

We evaluated self-reported pain experienced in the last month through VAS, depressive symptoms through PHQ-9 questionnaire and sleep quality through PSQI at baseline (T0), after one month/four acupuncture sessions (T1) and at the end of the seven months of treatment (T2). Blood samples were also collected for evaluation of inflammatory profile.

Acupuncture had a significant effect on chronic pain after 7-month treatment (VAS 6.8 ± 1.4 vs 3.5±1.8, p 0.001). An associated reduction of pro-inflammatory biomarkers TNF-α (9.13 ± 3.53 pg/mL vs 7.48 ± 3.27 pg/mL, p 0.037) and TREM-1 (587±299 pg/mL vs 473±237 pg/mL, p 0.001) was observed at the end of treatment. Depressive symptoms (8.4±4.9 vs 5.7±4.6, p 0.001) and sleep disorders (8.4 ± 4.1 vs 6.2 ± 3.3, p 0.004) improved after one month. All these effects persisted after 7 months of acupuncture.

The acupuncture treatment showed positive effects on chronic pain, probably mediated by an anti-inflammatory effect. Moreover, it determined improvement in quality of life after one month and then confirmed until the end of the treatment.

Keywords: *acupuncture, systemic autoimmune diseases, inflammation, autonomic nervous system, chronic pain*

[Abstract:1349]

REGARDING A CLINICAL CASE OF VASCULITIS ASSOCIATED WITH ANCA-MPO

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[1] ⚠ The authors did not provide affiliations upon requests from the event organizer

Introduction: Vasculitis associated with neutrophil cytoplasmic autoantibodies (ANCA) are a group of disorders that include microscopic polyangiitis (PAM). PAM predominantly affects small arteries and presents characteristics similar to other vasculitis in this group, making its diagnosis challenging.

Clinical Case: A 69-year-old woman, diagnosed with high blood pressure at age 50, with no other relevant history was admitted to the emergency department (ED) due to constitutional syndrome characterized by anorexia, asthenia and weight loss of 6 kg over 1 month. Associated with symmetric poly-arthralgias with an inflammatory rhythm and the appearance of punctate erythematous skin lesions located on the hands and feet, as well as confluent purplish lesions on the right lower limb. On objective examination, a purpuric lesion measuring approximately 4 cm on the right lower limb and an ulcerated lesion with necrotic tissue on the medial ⅓ of the left lower limb. From the etiological study, biopsy of skin lesions revealed neutrophilic vasculitis with damage to the vessel walls - leukocytoclastic vasculitis. In the thoracic study, fibrotic streaks with a residual appearance on the lung vertices. Analytically, normal renal function, with hematoproteinuria with 921 mg of proteins in 24 hours; normocytic, normochromic anemia with SV of 98 mm/h, without eosinophilia. The ANCA autoimmune study was positive with a titer of 1/640, MPO. He started methylprednisolone pulses with significant clinical improvement of his complaints and purpura resolution.

Discussion: The present clinical case demonstrates the importance of taking clinical history, physical examination in patients with a high index of suspicion for vasculitic syndrome.

Keywords: *vasculitis, ANCA-MPO, purple*

[Abstract:1372]

YOUNG WOMAN WITH MONOCLONAL GAMMOPATHY OF RENAL SIGNIFICANCE AND SJOGREN'S SYNDROME

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Case Report: We report the case of a 40-year-old woman with a child born full-term and having had with non-Hodgkin B MALT lymphoma in the right parotid gland in 2010, with complete remission.

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In August 2021, she was referred to our consultation for skin lesions in hel lower limbs compatible with purpura, arthralgia mainly in hands, angioedema and photosensitivity with rheumatoid factor 1100 (IU/ml), positive ANA for 1/80, antibodies anti SS-A 644 (U/ml) and anti SS-B 136 (U/ml) with consumption of C3 and C4.
At the anamnesis the patient referred dry mouth and eyes, which started during the treatment of lymphoma and which she attributed for years to a side effect of radiotherapy.
The patient was diagnosed with primary Sjogren syndrome and started treatment.
During follow-up, due to recurrence of purpura with no response to treatment, we performed a skin biopsy finding small secondary vessel vasculitis. In one of the purpura outbreaks the patient had also proteinuria in nephrotic range and type II mixed cryoglobulinemia so renal biopsy was performed finding membranoproliferative glomerulonephritis with kappa plasma cell infiltration.
In first instance we started rituximab but it had no effect.
The case was then presented at an interdisciplinary committee including pathological anatomy, rheumatology, nephrology and autoimmune diseases, where we concluded that the patient presented a monoclonal gammopathy of renal significance (MGRS).
After this diagnosis, treatment with bortezomib was started with good response.
Conclusions: - Lymphoproliferative syndromes and cryoglobulinemic vasculitis may be the initial presentation form of Sjogren syndrome.
- MGRS requires a high degree of suspicion and quick diagnosis and treatment to avoid irreversible renal impairment.

Keywords: *monoclonal gammopathy of renal significance, Sjogren's syndrome, cryglobulin*

[Abstract:1400]

DIFFICULTY IN DIAGNOSING GRANULOMATOSIS WITH POLYANGIITIS WITHOUT RENAL INVOLVEMENT

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Background: Granulomatosis with polyangiitis (GPA, previously named Wegener's) is a rare vasculitis with about 10 cases per million, which can be even more difficult to diagnose without renal injury. We aim to shed light on the other clinical manifestations and classification criteria of GPA besides pulmonary-renal syndrome.
Case Description: We present the case of a 51-year-old man who had a six-month history of weight loss, night sweats, nasal surgery for chronic congestion in addition to bilateral sensorineural hearing loss which required ear tube insertions. A thorax CT showed an incidental 3cm mass; initially misdiagnosed as a tumor, the lung nodule was biopsied twice only to have both pathology reports as dense fibrosis with lymphoplasmacytic cell infiltration and adipose tissue but no malignant cells. PET CT suggested inflammatory activity, and he was undiagnosed for months. The patient was admitted to our hospital with acute kidney injury, anemia and back pain with an initial hypothesis as multiple myeloma. He complained of polyarthralgia of large joints and redness of both eyes. Upon an ophthalmology examination, he was determined to have episcleritis. The addition of rheumatologic complaints led to a positive c-ANCA (PR3-ANCA), finally being diagnosed as granulomatosis with polyangiitis (Wegener's).
Discussion: GPA necrotizes small blood vessels, particularly impacting the lung and kidney that can lead to rapidly progressive glomerulonephritis or pulmonary hemorrhages, both of which have high mortality rates if left untreated.

REFERENCE:
Lutalo PMK, D'Cruz DP. Diagnosis and classification of granulomatosis with polyangiitis (aka Wegener's granulomatosis). Journal of Autoimmunity 2014 (48-49):p94-98.

Keywords: *GPA, vasculitis, granulomatosis with polyangiitis, Wegener's*

[Abstract:1437]

NOT SO MUCH OF A CHILDREN'S THING

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We present the case of a 64-year-old woman who presented to the emergency department with several days of evolving abdominal pain, primarily localized in the right hemiabdomen. Concurrently, she reported the appearance of non-painful and non-pruritic purpuric lesions on both thighs and diffuse arthralgia without associated inflammation. Upon arrival, the patient was in poor general condition. Physical examination revealed a painful abdomen with signs of peritonism. Purpura was palpable on the anterior aspect of both thighs, displaying lesions at different evolutionary stages. Laboratory findings showed worsening renal function, with a creatinine level of 2.2 mg/dL, and elevated acute phase reactants. Urinalysis revealed abundant red blood cells and few leukocytes. An abdominal CT scan was performed, revealing signs suggestive of regional peritonitis along with free intraperitoneal fluid. Paracentesis yielded serohematic fluid with parameters consistent with secondary bacterial peritonitis. Urgent surgical intervention was ruled out by General Surgery. Biopsy of the skin lesions was performed. However, the patient had a torpid evolution in the subsequent days and succumbed 48 hours later.
Subsequently, skin biopsy results revealed leukocytoclastic vasculitis consistent with the IgA type.
Despite being the most common systemic vasculitis in children with a benign course, IgA vasculitis is considered a rare entity

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in adults with a gloomier prognosis. Peritonitis is a serious and uncommon complication, secondary to involvement of mesenteric vessels. Regarding treatment, there are studies supporting the use of corticosteroids and other immunosuppressants in severe cases. However, their role is not clearly established, and their use remains controversial.

Keywords: *purpura, peritonitis, Iga vasculitis,*

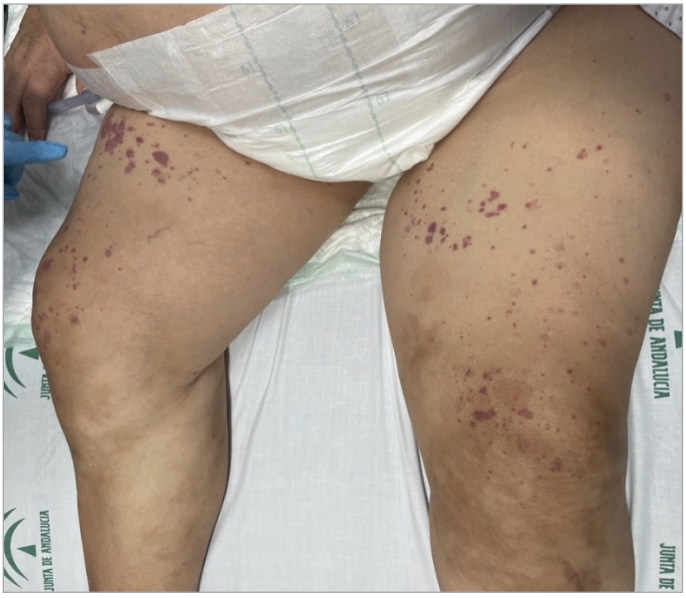


Figure 1. *Skin purpura of both patient's legs.*

[Abstract:1454]

ADULT CELIAC DISEASE PRESENTED WITH CELIAC CRISIS: REPORT OF TWO CASES

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Background: Celiac crisis is a rare presentation of celiac disease characterized by acute, severe metabolic imbalances resulting in high mortality and morbidity, with severe diarrhea, hypoproteinemia, metabolic and electrolyte disturbances.

Case Presentation: Two patients (case 1: 29 years old and case 2: 66 years old, female), with no known medical history of chronic diseases, presented to the hospital with prolonged diarrhea, weight loss, and severe hypocalcemia at different times.

They were admitted for hemodynamic instability in the setting of severe dehydration and electrolyte disturbances. Physical examination revealed a positive Trousseau sign in Case 1. The common laboratory features of both cases were low magnesium, low potassium, low vitamin D, low ferritin, and prolonged coagulation tests. In addition to those labs, case 2, had also metabolic acidosis. In both cases, the titers of the tissue transglutaminase IgA and IgG and the anti-endomysium antibody were high, and the histopathologies of the duodenal biopsy were consistent with villous atrophy, crypt hyperplasia, and an increase in intraepithelial lymphocytes, suggesting celiac disease. Both cases responded quickly to treatment with a gluten-free diet, fluid, electrolyte, vitamin D, and K replacements and were discharged.

Conclusions: Celiac crisis is typically seen in children under 2 years of age but can also be encountered in adulthood. Most cases respond to gluten cessation, nutritional support, and rarely steroid treatment.

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disease. Gastroenterology 2021;160(1):63-75.
2.Andersen DH, di'SantAgnese PA. Idiopathic celiac disease. I. mode of onset and
diagnosis. Pediatrics 1953;11(3):207-223.

Keywords: *celiac crisis, dehydration, diarrhea, electrolyte imbalance, metabolic disturbance*

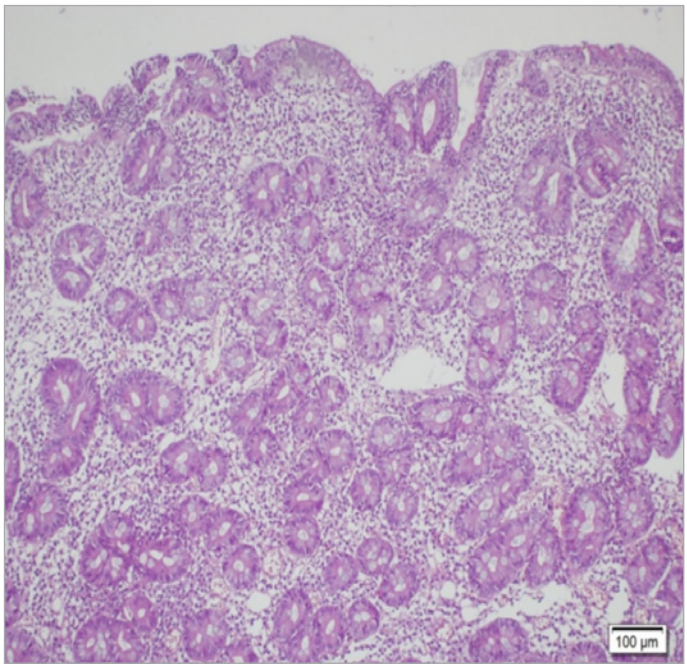


Figure 1. *Case 1: Duodenal biopsy image.*

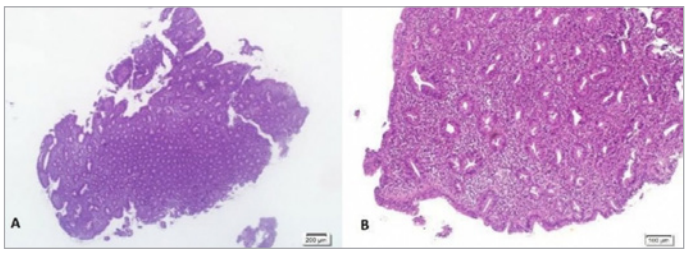


Figure 2. *Case 2: Duodenal biopsy image.*

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[Abstract:1455]

A RARE MANIFESTATION OF IG A VASCULITIS: PLEURAL EFFUSION

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Introduction: IgA vasculitis is an immunoglobulin A-mediated, small vessel vasculitis frequently affects children and young adults. The main tetrad of IgAV is: non-thrombocytopenic palpable purpura, abdominal pain, haematuria and arthritis/arthralgia. Pleural effusion is a rarely seen systemic manifestation in patients with IgAV. Herein we present a case with IgAV with a pleural effusion in the presentation.

Case: Our case is a 17-year-old female patient, without any known chronic disease, initially presented to our clinic with diffuse palpable purpuric rash, oedema in bilateral hands and feet, arthralgia, colicky abdominal pain and dyspnea. According to blood tests her ESR and CRP were found to be elevated and ANA test was positive. All remaining tests were in normal range. Her chest x-ray revealed pleural effusion and in abdominal imaging ileal wall thickening and oedema were detected accompanied with peritoneal fluid. Punch biopsy was taken from the skin lesions and resulted in leukocytoclastic vasculitis. Along with, colonoscopic biopsy from ileum was consisted with chronic ileitis. Corticosteroid was commenced and within 2 weeks, her rashes were dramatically regressed additionally, pleural and peritoneal effusions were disappeared. Corticosteroid regimen has been gradually decreased without any flare up.

Conclusions: In IgAV, non-blanching palpable purpuric rash, arthritis, abdominal pain and vomiting, subcutaneous oedema are the usual symptoms. Serositis are rarely seen, there are few reports with pleural involvement. We would like to highlight this infrequent manifestation of IgAV and share this educational case which has both intestinal and pleural involvement.

Keywords: *IgA vasculitis, pleural effusion, serositis*



Figure 1. *The initial presentation of the patient with disseminated palpable purpuric rashes.*

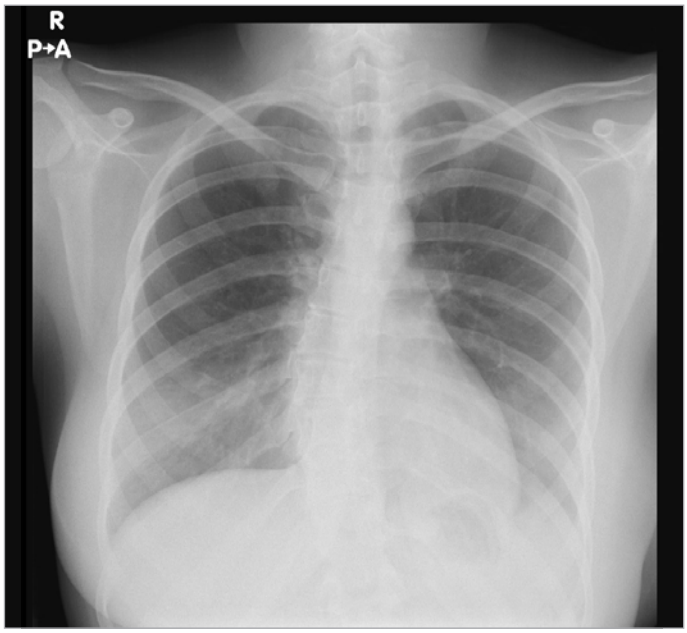


Figure 2. *The Chest X-Ray consisted with left pleural effusion.*

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Figure 3. Four weeks after the initiation of corticosteroids.

[Abstract:1469]

A CASE OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS, CHALLENGES IN DIFFERENTIAL DIAGNOSIS, ETIOLOGY, AND TREATMENT

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Hemophagocytic lymph histiocytosis (HLH) is characterized by tissue damage resulting from the excessive activation of macrophages. Infections, connective tissue diseases, and malignancies may be the trigger.

A 60-year-old woman applied to our clinic with fatigue. She had no history of chronic diseases or drug use before.

On examination, her body temperature was 38.3°C, and there were signs of mild hypervolemia. Laboratory tests showed normocytic anemia, thrombocytopenia, low fibrinogen, and extremely high ferritin levels. Biochemical tests showed acute renal injury, hypoalbuminemia, and elevated LDH and triglycerides, and the urine examination showed a nephrotic proteinuria of 18,600 mg/g (Laboratory findings are shown in Table 1). Schistocytes were less than 1% in the peripheral blood smear. Normal cell morphology and abundant hemophagocytic activity were observed in the bone marrow examination. The patient was diagnosed with HLH.

PCR panels performed for viral infections were negative. Bacterial and tuberculosis cultures, and other infectious serologic tests were negative. PET-CT was negative for neoplasia. ANA was found in 1/100-1/320 titres and speckled staining pattern, and other serologic rheumatologic tests were negative. Kidney biopsy showed AA amyloidosis. The patient was given corticosteroid, IVIG, and anakinra treatments, and recovered completely. FMF genetic testing was negative. Whole exome analysis showed no primary HLH associated genes, but TLR7 mutation, which can be related with systemic lupus erythematosus, and immunodeficiencies according to the genetic database.

The patient's AA amyloidosis, secondary HLH, and genetic findings were evaluated together, and continuation of the anakinra treatment was decided due to possible connective tissue disease.

Keywords: *hemophagocytic lymphohistiocytosis, AA amyloidosis, TLR7 mutation, anakinra*

Hemoglobin (12-14.9 g/dl)	10.7
White blood cell count (4.5-12.6x10^3/uL)	13.4
Platelet count (175-390x10^3/uL)	48
Fibrinogen (200-400 mg/dl)	74.5
D-dimer (0-0.5 mg/L)	52.72
Eryhtrocyte sedimentation rate (0-20 mm/h)	18
CRP (0-5 mg/L)	79.2
BUN/Creatinine (8-23/0.5-0.9 mg/dl)	77/5.8
Sodium/Potassium (135-145/3.5-5.1 mEq/L)	136/3.8
Calcium/Phosphorus n(8.6-10.2/2.7-4.5 mg/dl)	7.3/6.5
Protein/Albumin (6.4-8.3/3.5-5.2 g/dl)	5.9/2.9
LDH(135-214 U/L)	691
AST/ALT (0-31/0-33 U/L)	21/17
ALP/GGT (0-64 U/L / 6-42 IU/L)	88/59
Total/direct bilirubin (0-1.1/0-0.2 mg/dl)	0.36/0.22
Triglycerides (57-149 mg/dl)	272
LDL-cholesterol (100-129 mg/dl)	68
Ferritin (13-150 ng/ml)	30795

Table 1. Laboratory findings on admission.

[Abstract:1480]

A CASE OF SEVERE MACROPHAGE ACTIVATION SYNDROME IN ADULT PATIENT SUCCESSFULLY TREATED WITH INTERLEUKIN-1 INHIBITOR

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Case Report: Macrophage Activation Syndrome (MAS) is life-threatening syndrome massive cytokine storm triggered by an autoimmune systemic disease (1).

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A 48 years old woman with a history of Systemic Lupus Erythematosus presented with fever, fatigue, hyporexia, vomit, jaundice and confusion. Blood exams showed pancytopenia with absolute neutropenia, hyperferritinemia, increase of c-reactive protein, direct bilirubin, gamma-GT, transaminases and hypertriglyceridemia (not previously known).

Major viral, bacterial and mycotic infections were ruled out through extensive radiological, cultural, serological, and PCR amplification tests.

MAS was thus considered. The patients started high-dose corticosteroid therapy with no significant benefit. An interleukine-1 inhibitor, anakinra, was thus introduced with subsequent rapid clinical improvement. We initially, started at 100 mg IV/6 hours then tapered to 100 mg/8 hours after clinical response. In the following days CRP, ferritinemia and hypertriglyceridemia, total bilirubin and cytopenia improved, but after 14 days of therapy, a drop in neutrophil count and elevation of total bilirubin was observed. Anakinra was thus tapered to 100 mg/day and cyclosporine (50 mg twice daily) added with clinical and laboratory improvement. The patient was finally discharged.

Conclusions: Although not specifically approved in the adult, a recent position paper has suggested its use (1) and there is evidence on its benefit in pediatric patients (2). The case is one of the few reports on the use of a IL-1 inhibitor anakinra as an effective treatment in severe MAS.

Reference:
1] Hines, M. R. et al. 20222] Griffin, G. et al. 2020

Keywords: *macrophage activation syndrome, anakinra, adult patient*

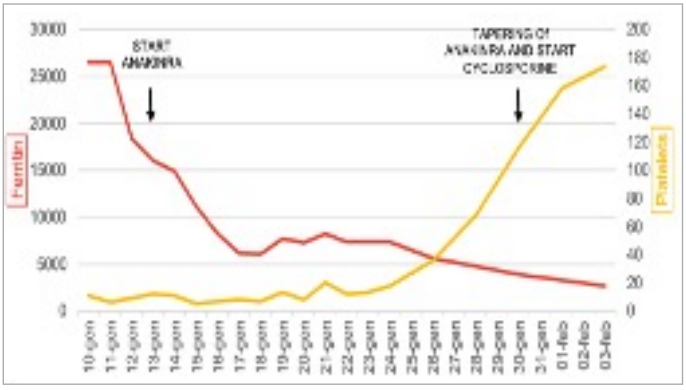


Figure 1. *Time course of ferritin and platelet's count.*

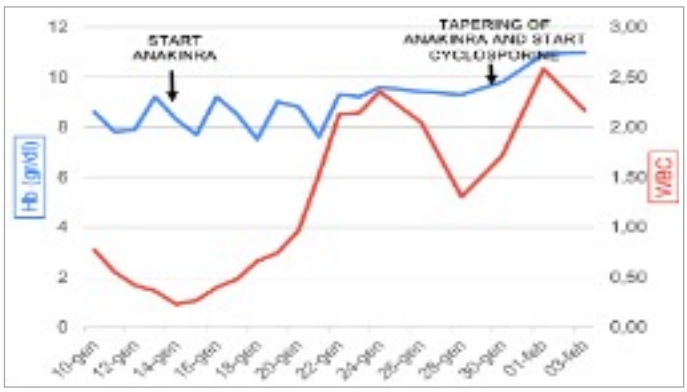


Figure 2. *Time course of hemoglobin and white blood cells.*

[Abstract:1483]

CARDIOGENIC SHOCK IN A 16-YEAR-OLD GIRL

Salvador Aguilar Alba, Ana Isabel Pérez Caballero, Laura Limia Pérez, Aquiles Lozano Rodríguez Mancheño, Juan Criado García, José López Miranda

Lipids and Atherosclerosis Unit, Department of Internal Medicine, Maimonides Biomedical Research Institute of Cordoba (IMIBIC), Reina Sofia University Hospital, University of Cordoba, Cordoba, Spain

We present a 16-year-old female with a history of headache, asthenia and weakness in the lower limbs for 4 months, who attended the emergency department for dyspnoea. She presented pulmonary infiltrates, NT-proBNP 130,000 pg/mL, troponins 10,810 ng/L and C-reactive protein 42 mg/L, together with severe respiratory failure and the need for intubation in the intensive care unit (ICU).

Echocardioscopy showed severe aortic insufficiency, global akinesia and the angio-computed tomography (angio-TC) revealed a infrarenal aortitis, thrombi in the ascending aorta and pulmonary emboli (Figure 1).

Suspicion of large vessel vasculitis led to treatment with boluses of methylprednisolone 500 mg for 5 days with a subsequent decrease to a dose of mg/kg/day. Due to haemodynamic instability, a multidisciplinary committee decided on surgical intervention with placement of a valved stent. The surgical procedure was not satisfactory, and the patient was left with Veno-Arterial-ECMO and haemofiltration due to renal failure.

At 48 hours, he had an areactive anisocoria as a consequence of a malignant right middle stroke. In the following days she had bad prognosis. On day +21 of admission to the ICU, tocilizumab 8mg/kg/single dose was administered. The following day she again presented intracranial bleeding. On day +23 a coronary-CT and angio-CT of the aorta confirmed critical lesions of both coronary arteries and complete stenosis of the abdominal aorta.

Finally, the patient died, and a clinical autopsy was performed which confirmed the ischaemic damage in the macroscopic specimen and the pathological anatomy showed the typical findings of Takayasu's arteritis (Figure 2).

 DOI: 10.12890/2024_V11Sup1

Keywords: *aortic insufficiency, aortitis, Takayasu's arteritis, tocilizumab*

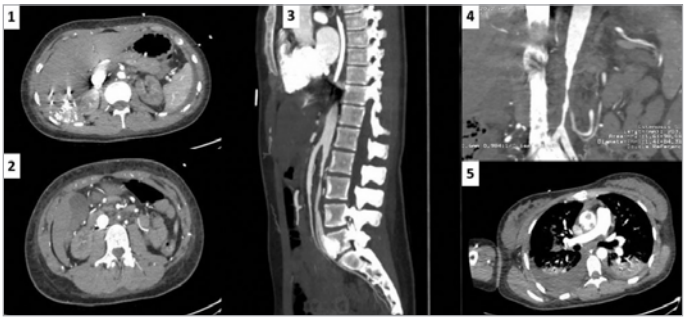


Figure 1. *Angio-CT of the aorta performed on the patient's admission to the ICU.*
1. Axial section showing the normal calibre aortic artery before the exit of the renal artery. 2. Very marked decrease in the calibre of the aortic artery can be seen below the renal artery outflow. 3. Sagittal view showing the decrease in the calibre of the infrarenal abdominal aorta from a different perspective. 4. Coronal view of the infrarenal abdominal aorta. 5. Three endoluminal repletion defects are shown 1 cm from the sinotubular junction of the ascending aorta, very close to the aortic valve, which are suggestive of thrombus.

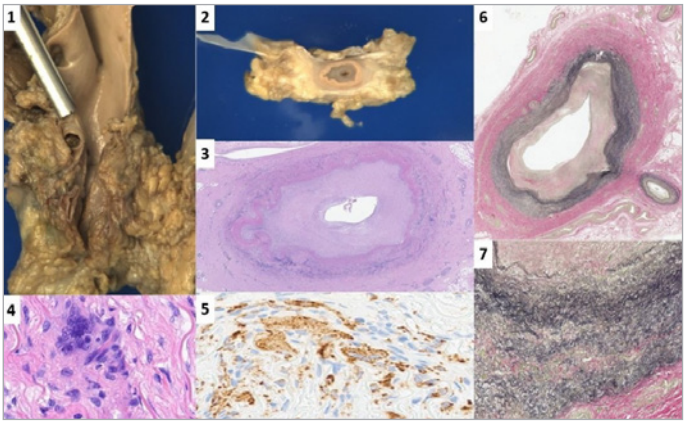


Figure 2. *Clinical autopsy images of the patient.*
1. The macroscopic specimen shows a significant reduction of the abdominal aorta below the celiac trunk affecting the renal arteries, common iliac artery, external and common femoral arteries. 2. Cross section showing thickening of the medial layer with irregularly reduced calibre of the arterial lumen giving a filiform appearance. 3. Histological section with segment of aortic artery with intense involvement of the media due to chronic inflammatory component. 4. Presence of multinucleated giant cells in the foci of media destruction. 5. CD3+ labelling. 6-7. Myointimal thickening with rupture of elastics and coagulation phenomena with quasi-critical reduction of the artery.

[Abstract:1503]

BONE MINERAL DENSITY AND SERUM NETRIN-1 LEVELS IN POSTMENOPAUSAL OSTEOPOROSIS EVALUATION OF THE RELATIONSHIP

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[1] Department of Internal Medicine, Ankara City Hospital, Ministry of Health, Ankara, Turkey
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Netrin-1 has been shown to be a bone-related molecule. It was thought that plasma Netrin-1 level could be a biomarker that can be used for diagnosis and evaluation of disease activity in postmenopausal osteoporosis patients. 59 volunteer patients with osteoporosis over the age of 40 who applied to Ankara Bilkent City Hospital Internal Medicine outpatient clinic, 54 osteopenia and 63 volunteer healthy individuals, were included. No significant difference was detected between the groups in terms of serum Netrin-1. No significant difference was found between the groups and the ages and heights of the patients. Body weight in the osteoporosis group was significantly lower than the osteopenia and control groups. MI was found to be significantly lower in the osteoporosis group compared to the osteopenia and control groups. No significant difference was detected between serum osteocalcin levels of groups. The measured calcium values of the patients did not differ significantly between the groups. Spearman correlation analysis was performed to determine the relationship between serum Netrin-1 levels and some biochemical parameters. No significant correlation was observed between serum Netrin-1 level and T score, osteocalcin, vitamin D and BMI values measured by DEXA method. No significant correlation was observed between the T score measured by the DEXA method and osteocalcin and vitamin D values. A significant negative correlation was found between the T score measured by the DEXA method and BMI. No correlation was observed between serum osteocalcin levels, vitamin D and BMI. A significant negative correlation was detected between vitamin D and BMI. Netrin-1 molecule, which is accepted as a bone mineral marker, is a scoring system used to evaluate disease activity scores.

Keywords: *postmenopausal osteoporosis, bone mineral density, Netrin-1*

 DOI: 10.12890/2024_V11Sup1

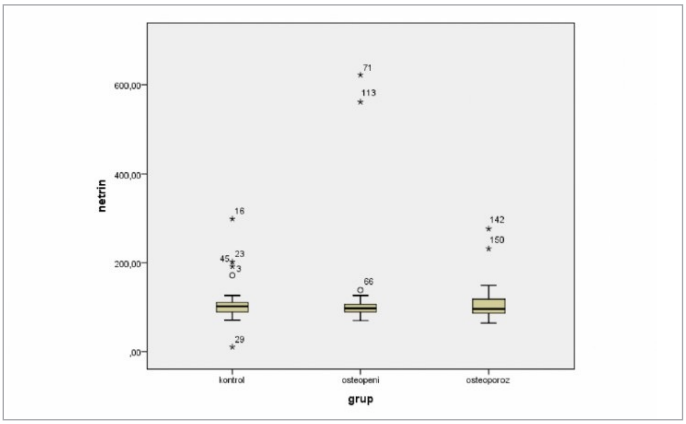


Figure 1. *Box Plot distribution of serum Netrin-1 levels in osteoporosis, osteopenia and control groups.*

Box Plot distribution of serum Netrin-1 levels in osteoporosis, osteopenia and control groups is shown in Figure 1, and Netrin-1 median values were found to be similar between the groups.

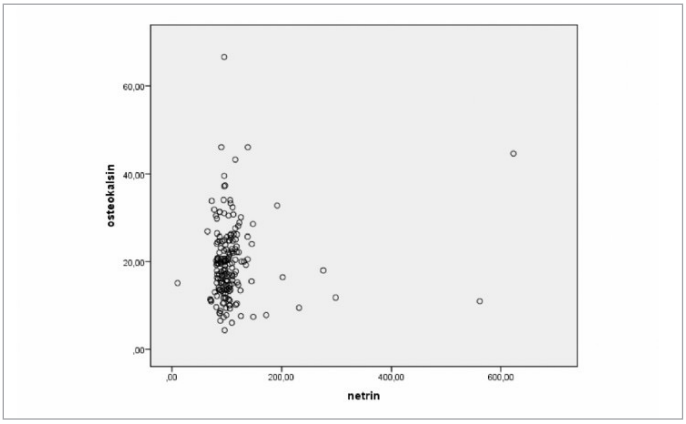


Figure 2. *Distribution of Netrin-1 levels according to osteocalcin levels in the Scattering graph.*

The distribution of Netrin-1 levels according to osteocalcin levels is shown and no significant correlation relationship was detected between osteocalcin levels and Netrin-1 levels.

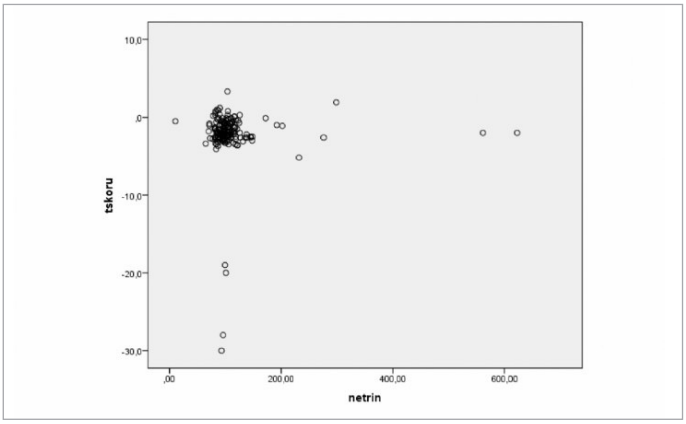


Figure 3. *The distribution of Netrin-1 levels according to T scores is shown in the Scattering graph.*

	Control (n=63)	Osteopenia (n=54)	Osteoporosis (n=59)	P-Value
DEXA T score, median (IQR)	-0.5 (-0.9- [-0.1])	2.0 (-2.2- [-1.7])	2.8 (-3.3- [-2.6])	<0.001
Netrin-1, median (IQR) [pg/mL]	102.49 (89.73-111.35)	98.09 (89.66-106.85)	96.13 (86.85-118.47)	0.719
Osteocalcin, median (IQR) [ng/mL]	17.06 (13.22-22.90)	19.76 (13.66-24.29)	19.62 (15.49-25.70)	0.141

Table 1. *Comparison of DEXA T score, Netrin-1 and Osteocalcin values between groups.*

The median value of the Lumbar T score measured by the DEXA method in the osteoporosis group was -2.8, and the median value in the osteopenia group was -2.0, median value in the normal group; It was found to be -0.5. When serum Netrin-1 levels were evaluated between the groups, the median Netrin-1 value was found to be 96.13 pg/mL in the osteoporosis group, 98.09 pg/mL in the osteopenia group and 102.49 in the control group. When the results were evaluated, no significant difference was found between serum Netrin-1 levels obtained by ELISA method in the osteoporosis, osteopenia and control groups (p value 0.719). When serum osteocalcin levels were evaluated between the groups, the median osteocalcin value was found to be 19.62 ng/mL in the osteoporosis group, 19.76 ng/mL in the osteopenia group and 17.06 ng/mL in the control group. When the results were evaluated, there was no significant difference between the serum osteocalcin levels of the osteoporosis, osteopenia and control groups (p value 0.141).

	Control (n=63)	Osteopenia (n=54)	Osteoporoz (n=59)	P-Value
WBC, median (IQR)	6380 (5020-8170)	6360 (5415-7530)	5990 (5150-7000)	0.624
HGB, median (IQR)[g/dL]	13.3 (12.4-14.2)	13.7 (12.3-14.3)	13.0 (12.3-13.8)	0.145
Platelet, median (IQR) [x10^9/L]	242 (216-306)	256 (228.50-304.75)	270 (223-330)	0.435
TSH(thyroid stimulating hormone), median (IQR) (mU/L) [mU/L]	1.60 (1.22-2.57)	1.95 (1.43-3.03)	1.72 (1.00-2.54)	0.178
Calcium, median (IQR) [mg/dL]	9.6 (9.2-10.0)	9.6 (9.3-10.0)	9.5 (9.0-10.0)	0.316
Phosphorus median (IQR) [mg/dL]	3.6 (3.3-4.0)	3.7 (3.5-3.9)	3.6 (3.3-3.9)	0.631
Parathyroidhormone, median (IQR) [ng/L]	51.0 (42.0-65.0)	63.0 (47.8-78.5)	57.0 (45.0-79.0)	0.056
Vitamin D median (IQR)] [nmol/L]	45.0 (25.0-62.0)	44.0 (26.0-70.0)	49.0 (25.0-66.0)	0.685
ALP(alkaline phosphatase), median (IQR) [U/L]	82.0 (67.0-100.0)	80.0 (698-96.5)	88.0 (68.0-102.0)	0.643
LDH(lactatedehydrogenase), median (IQR) [U/L]	208.0 (187.0-226.0)	211.5 (189.0-240.0)	205.0 (186.0-230.0)	0.671
Albumin, median (IQR) [g/L]	45 (43-47)	45 (43-47)	44 (41-47)	0.164

Table 2. *Comparison of other laboratory data between groups.*

The median calcium value was found to be 9.6 mg/dl in the control group, 9.6 mg/dl in the osteopenia group, and 9.5 in the osteoporosis group. There was no significant difference in the measured calcium values of the patients between the osteoporosis, osteopenia and control groups (p value 0.316). The median value of vitamin D was found to be 45 mg/dL in the control group, 44 mg/dL in the osteopenia group, and 49 in the osteoporosis group. There was no significant difference in the measured vitamin D values of the patients between the osteoporosis, osteopenia and control groups (p value 0.685). Among the hemogram parameters of the patients participating in the study, WBC, HGB, Platelet; TSH, phosphorus, PTH, ALP, LDH, Albumin laboratory values were examined and no statistically significant difference was found between the groups (p>0.005).

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r (p)	Netrin-1	DEXA T score	Osteocalcin	Vitamin D	BMI
Netrin-1		-0.007 (0.931)	0.074 (0.331)	0.004 (0.959)	0.005 (0.945)
DEXA T score			-0.140 (0.065)	-0.033 (0.668)	-0.249 (0.001)**
Osteocalcin				-0.096 (0.207)	-0.035 (0.643)
Vitamin D					-0.160 (0.034)*

Table 3. *Correlation analysis results between some operating parameters.*

Serum Netrin-1 düzeyleri ile biyokimyasal bazi parametreler arasindaki ilişkiyi belirlemek üzere Spearman korelasyon analizi yapildi. Yapilan bu analizler Tablo 7'de gorulmektedir. Serum Netrin-1 duzeyi ile, DEXA yontemiyle olculen Lomber T skoru, osteokalsin, D vitamini ve VKI değerleri arasinda anlamli korelasyon gözlenmedi (sirasiyla r =0.007, p =0.931 ve r =0.074, p =0.331 ve r =0.004, p=0.959 ve r=0.005, p=0.945). DEXA yontemi ile olculen Lomber T skoru ile, osteokalsin ve D vitamini deArial erleri arasinda anlamli bir korelasyon gözlenmedi (sirasiyla r = -0.140, p=0.065 ve r = -0.033, p= 0.668). DEXA yontemi ile olculen Lomber T skoru ile VKI arasinda negatif yonde anlamli korelasyon saptanmistir (sirasiyla r = -0.249, p=0.001). Serum osteokalsin duzeyleri ile, D vitamini ve VKI arasinda korelasyon gozlenmedi (sirasiyla r = -0.096, p = 0.207 ve r = -0.035, p=0.643). D vitamini ile VKI arasinda negatif yonde anlamli korelasyon saptandi (sirasiyla r = -0.160, p=0.034).

⚠ *The authors did not provide an English version of Table 3 caption upon requests from the event organizer.*

	Control (n=63)	Osteopenia (n=54)	Osteoporosis (n=59)	P-value
Age, median (IQR) (years)	59.0 (53.0-67.0)	61.0 (57-69.3)	62.0 (57.0-70.0)	0.294
Height, median (IQR)(cm)	159.0 (155.0-162.0)	160.0 (155.0-165.0)	157.0 (154.0-161.0)	0076
Body weight, median (IQR)(kg)	73.0 (65.0-83.0)	69.5 (63.0-77.5)	65.0 (60.0-71.0)	0.001*
BMI, median (IQR)(kg/m²)	29.24 (25.97-33.06)	27.78 (24.02-30.40)	26.08 (23.13-29.09)	0.004*

Table 4. *Meeting demographic characteristics across groups.*

Demographic characteristic data were compared between groups. Among the patients included in our study, participants with a T score ≥ -1 SD in BMD measurements were included in the control group (n = 63); Participants with a T score between -1 and -2.5 were included in the osteopenia group (n = 54), and participants with a T score ≤ -2.5 were included in the osteoporosis group (n = 59). Comparison of demographic parameters of the study groups can be seen in Table 4. The median age was found to be 62 in the osteoporosis group, 61 in the osteopenia group, and 59 in the normal group. The median age value in the osteoporosis group was found to be higher than the osteopenia and control groups, but this height was not statistically significant (p value 0.294). The median height value was found to be 157 cm in the osteoporosis group, 160 cm in the osteopenia group, and 159 cm in the control group. There was no statistically significant difference in the height of the patients between the osteoporosis, osteopenia and control groups (p value 0.076). The median value of body weight in the osteoporosis group was 65 kg, in the osteopenia group it was 69.5 kg, and in the control group it was 73 kg. Body weight was found to be statistically significantly lower in the osteoporosis group compared to the osteopenia and normal groups (p value 0.001). The median BMI value was found to be 26.08 kg/m² in the osteoporosis group, 27.78 kg/m² in the osteopenia group, and 29.24 kg/m² in the control group. Statistically, BMI was found to be significantly lower in the osteoporosis group compared to the osteopenia and normal groups (p value 0.004).

[Abstract:1535]

BALANCE DISORDERS AND RISK OF FALLS IN RHEUMATOID ARTHRITIS

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Introduction: Patients with rheumatoid arthritis (RA) are exposed to an increased risk of balance disorders and falls due to chronic inflammation that affects joints and surrounding tissues.

These balance disorders are multifactorial and result from the interaction of several mechanisms:

stiffness alters the walking pattern, muscle weakness and pain can affect the ability to maintain a stable posture and make rapid balance adjustments in response to disturbances.

Visual impairments, psychiatric manifestations (depression, anxiety) can also impact patients' balance, kinesiophobia, can further affect balance.

Specific treatments can also be implicated, comorbidities increase the risk of falls, particularly in patients also suffering from osteoporosis.

Screening for balance disorders and patients at risk of falls needs for a comprehensive evaluation.

Several validated balance assessment tools enable the detection of patients with disorders and, consequently, at risk of falls. These tools also assess the effectiveness of prevention interventions.

Role of Physical Therapy: It is essential to improve the quality of life and reduce associated healthcare costs, it must be personalized and tailored to the specific needs of each patient.

Physical therapy unit can collaborate with psychologists, neuropsychologists, and social workers to help patients manage the psychological and social aspects of the disease.

Non-pharmacological interventions are used to improve muscle strength, proprioception, postural stability, and reduce kinesiophobia.

Conclusions: Balance disorders with the risk of falls are common in patients with RA but often overlooked. Evaluation should involve validated tools. Early and proactive management is essential to improve the quality of life for these patients and prevent complications.

Keywords: *rheumatoid arthritis, balance, falls*

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[Abstract:1559]

DON'T FORGET THE DRUGS. AN ATYPICAL CASE OF RENOPULMONARY SYNDROME IN AN ADULT MALE

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Case Presentation: We report the case of a 69-year-old male, ex-smoker and alcoholic, treated with oral allopurinol for hyperuricemia and gouty crises, living independently in a rural area. He initially presented to the emergency department with dyspnoea, astenia and fever and was diagnosed with community-acquired pneumonia. Despite initial treatment, he returned within a week with persistent symptoms. Blood tests revealed anaemia, elevated inflammatory markers and signs of renal impairment. Imaging studies showed bilateral pulmonary infiltrates and progressive deterioration of renal function, accompanied by hypertension.

Hypothesis: Given the clinical presentation and the consideration of a renal-pulmonary syndrome, the differential diagnosis focuses on primary ANCA-associated vasculitis versus drug-induced ANCA-associated vasculitis, especially in association with allopurinol.

Diagnostic Pathways: Serial blood cultures, IGRA for M. tuberculosis, sputum and mycobacteria culture, sputum and bronchoaspirate smear microscopy were performed with negative results. Infectious serologies were extracted without relevant findings. Proteinogram, lymphocyte subpopulations, immunoglobulins and light chains were normal. Autoimmunity panel was requested with positivity to low titers of ANCA of perinuclear profile MPO1, with negativity for antinuclear antibodies and extractable anti-nuclear antigen. Finally, a renal biopsy was performed showing ANCA-associated glomerulonephritis (MPO) with accompanying deposits of immunocomplexes.

Discussion and Learning Points: Clinical judgement leans towards primary or drug-induced ANCA-associated vasculitis, emphasising the importance of considering these entities in the context of renal-pulmonary syndromes. Recognition of drug-induced vasculitis is critical and has implications for prognosis and treatment. Early identification and withdrawal of the offending agent have a significant impact on management and differentiate it from primary vasculitis.

Keywords: *vasculitis, allopurinol, renopulmonary syndrome, hyperuricemia*



Figure 1. *Posteroanterior chest readiography showing bilateral pulmonary infiltrates.*



Figure 2. *Thoracic CT scan reporting multiple bilateral peripheral consolidating pulmonary opacities suggestive of organized, nonspecific pneumonia.*

[Abstract:1573]

SUCH A COMMON VIRUS

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A 19-year-old woman referred arthralgias in both knees which evolved distally to the ankles with myalgias in the lower limbs at 48 hours of evolution, causing significant functional impotence. In the last 24h, arthralgia intensified affecting the upper limbs too. She denied fever or other accompanying symptoms.

Physical examination revealed that she was hemodynamically stable, with no remarkable findings except petechiae on the inner face of both thighs and isolated petechiae in the dorsal region.

Blood tests showed bilirubin elevation at the expense of the indirect fraction, the elevation of acute phase reactants, a decreased hemoglobin levels, thrombocytopenia and coagulopathy.

She was admitted to the Internal Medicine department with the main suspicion of hemolytic anemia, which was confirmed by the decrease in haptoglobin levels.

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To clarify the origin of this clinical picture, we corroborated the anamnesis and re-evaluated the patient, who had reported an episode of odynophagia two weeks earlier.
The study was extended with new complete laboratory tests, showing elevated lactate dehydrogenase, D-dimer and positive serology (IgM and IgG) for Epstein-Barr virus (EBV) with positive cryoglobulins. The rest of the autoimmunity, serologies and studies were negative.
With the diagnosis of hemolytic anemia and thrombopenia secondary to EBV infection, treatment was started with steroids and ceftriaxone for 5 days achieving normalization of both clinical and analytical parameters and the patient could be referred home.
Cryoglobulin-associated hemolytic anemia and thrombopenia are unusual manifestations, forcing us to rule out complications such as viral infections like EBV, hemophagocytic syndrome or lymphoproliferative processes.

Keywords: *hemolytic anemia, thrombopenia, Epstein-Barr, cryoglobulins.*

[Abstract:1575]

INFLAMMATORY MYOPATHIES - INSIGHTS FROM A MOROCCAN POPULATION

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Introduction: Inflammatory myopathies are rare autoimmune conditions characterized by inflammation of skeletal muscles and the skin. Their pathophysiology is still poorly understood.
Purpose: To determine the epidemiological, clinical, paraclinical, therapeutic, and progressive characteristics of inflammatory myopathies at the Mohammed VI University Hospital in Marrakech.
Findings: Forty patients were enrolled in the Department of Internal Medicine between August 2010 and January 2020. We identified 19 cases of dermatomyositis, 11 cases of polymyositis, and one case each of antisynthetase syndrome. The average age of our patients was 40 years, with a clear female predominance (F/M ≈ 5) and an average consultation delay of 15 months.
Clinical manifestations were predominantly muscle weakness and skin rash, found in 82.5% and 40% of patients, respectively. Rhabdomyolysis was observed in 77.5% of cases, with positive immunological findings in 66.6% of cases. A paraneoplastic origin was confirmed in three patients, involving breast tumors, Hodgkin's lymphoma, and small-cell lung carcinoma. All patients received corticosteroid therapy, with additional immunosuppressive treatment in cases of refractory disease (5% of cases). 85% of patients experienced partial remission, while 15% achieved complete remission.
Conclusions: Our results highlight diagnostic delays and challenges in establishing a clear prognosis for these patients. This underscores the importance of standardized management and appropriate care to prevent complications that can impact both vital and functional prognosis.

Keywords: *inflammatory myopathies, autoimmune diseases, antibodies*

[Abstract:1577]

PLATELETS AND SYSTEMIC LUPUS ERYTHEMATOSUS: VICTIM OR ACCOMPLICE?

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Introduction: Systemic lupus erythematosus (SLE) is a complex connective tissue disease with multisystemic involvement. Platelets were proposed as both contributors to its development and targets of autoimmune processes.
Purpose: To evaluate the frequency and characteristics of lupus patients with thrombocytopenia and to assess the role of thrombocytopenia as a risk factor in these patients.
Methods: This is a 13-year retrospective study (2010-2022) conducted on SLE patients at Mohammed VI University Hospital's Internal Medicine Department in Marrakech
Findings: Among the 342 SLE cases, 23.4% had thrombocytopenia (80 cases). The average age was 32.4 years, with a female predominance (F/M = 1/39). Thrombocytopenia was associated with higher SLE Disease Activity Index (SLEDAI) scores (11 vs. 9.6) and primarily linked to joint (71.8%, p=0.04) and dermatological (77.9%, p=0.3) involvement. Renal involvement was slightly more common in thrombocytopenic patients (31.1% vs. 25.3%). Thrombocytopenia occurred alone in 13.6% of cases, often with anemia (78.2%) and lymphopenia (58.2%, p=0.03). Anti-nuclear antibodies were positive in 94.4% (p=0.5) of thrombocytopenic patients, with 85.1% also having anti-DNA antibodies (p=0.03). Hydroxychloroquine was used in 80% of cases, and 92.5% received systemic corticosteroids. Immunosuppressants and biological treatments were considered, especially when hematological involvement was combined with other systemic issues, with two cases requiring rituximab for refractory thrombocytopenia.
Conclusions: Thrombocytopenia in SLE remains a fairly common condition. Its association in the literature with severe systemic involvement and high mortality makes it a warning sign to watch for in these patients to ensure better monitoring and improved prognosis.

Keywords: *systemic lupus erythematosus, thrombocytopenia, autoimmune diseases*

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[Abstract:1616]

LATE-ONSET LUPUS: CLINICAL FEATURES IN A MOROCCAN COHORT

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Introduction: Systemic lupus erythematosus (SLE) is a rare connective tissue disease, affecting primarily young women. Late-onset SLE is defined as when the diagnosis is made at the age of 50 and older. There is limited research on this particular subset of SLE.

Purpose: To evaluate the clinical, laboratory, and therapeutic characteristics of late-onset lupus within a Moroccan population.

Methods: This is a 13-year retrospective study (2010 - 2022) focusing on patients diagnosed with SLE after the age of 50 at CHU Mohammed VI's Internal Medicine Department in Marrakech.

Findings: Among 342 SLE cases, 39 (23.4%) had late-onset lupus, with an average age of 32.4 years, a notable female predominance (F/M=5/1) and an average diagnostic delay of 10.6 months, and concurrent autoimmune diseases in 5 cases. Upon admission, the mean SLE Disease Activity Index (SLEDAI) was 6.5. Cutaneous manifestations occurred in 94.9% of cases, including malar rash (58.3%) and photosensitivity 55.9%. Joint involvement was reported in 84.4% of cases, while renal, respiratory, and cardiac complications were noted in 33.3%, 30.8%, and 23.1% of cases, respectively. Laboratory findings revealed anemia in 62.5% (37.5% were hemolytic), lymphopenia in 44.7%, thrombocytopenia in 29.7%, positive antinuclear antibodies in 81.6%, and anti-native DNA antibodies in 63.3%. 88.6% of the patients received hydroxychloroquine, 94.4% were treated with systemic corticosteroids, while 43.6% received classic immunosuppressants. None of the patients received biologic therapies.

Conclusions: Late-onset SLE is rare condition, with distinct characteristics from younger individuals, reflecting the impact of age on the typical course of SLE.

Keywords: late-onset lupus, connective tissue diseases, autoimmune diseases

[Abstract:1618]

CLINICAL PATTERNS OF TAKAYASU ARTERITIS IN A MOROCCAN SETTING

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Introduction: Takayasu arteritis is a systemic inflammatory condition of unknown aetiology characterized by damage to the large and medium arteries and their branches, diagnosed typically through clinical suspicion and vascular imaging.

Purpose: To analyze the epidemiological, clinical, paraclinical, and therapeutic characteristics of this vasculitis in a moroccan population.

Methods: This is a descriptive retrospective study that includes patients treated for Takayasu arteritis in the Internal Medicine Department at Mohammed VI University Hospital in Marrakech from January 2010 to December 2020.

Findings: Twenty patients were included. The mean age was 39.7 years, with a clear female predominance (19F/1H). Claudication of the upper limbs was the chief complaint (30%), followed by ischemic stroke (20%), and other ischemic manifestations (15%). Although decreased visual acuity was reported in only 2 patients, ocular involvement was noted in 6 cases (30%). On clinical examination, peripheral pulselessness was found in 12 patients (60%), with vascular murmurs in 11 cases (55%). Biological inflammatory syndrome was reported in 14 patients, accounting for 70% of cases, with an average erythrocyte sedimentation rate of 51 mm. Nineteen patients received initial corticosteroid therapy, combined with methotrexate in 7 patients (35%) and cyclophosphamide in 2 patients. Tocilizumab was prescribed in 2 cases with incomplete responses to first-line treatment.

Conclusions: Takayasu arteritis is a rare but high-morbidity and high-mortality condition, with frequent relapses and progression of vascular lesions despite innovative treatments. Corticosteroids and methotrexate are the first-line treatment; subsequently, in cases of incomplete response, biologic therapies will be considered.

Keywords: Takayasu arteritis, large vessel vasculitis, internal medicine

[Abstract:1620]

UNDIFFERENTIATED AUTOINFLAMMATORY SYNDROME: A SMALL COHORT OF CASES

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Purpose: To describe a small and heterogeneous group of patients labeled with undifferentiated autoinflammatory syndrome.

Methods: This is a retrospective observational study developed in our clinic between 2014 and 2023, which includes patients with autoinflammatory syndromes without definitive confirmatory genetic studies.

Findings: Eight patients (4 men and 4 women) were included in this study, who had a mean delay time until diagnosis of 13.3 months. The main symptom was periodic fever (87% of patients)

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with an average number of 15.6 episodes per year and an average duration of 4.5 days. The second most frequent manifestation was arthralgia (50%) and in third place were myalgia and maculopapular rash (37.5%). Regarding treatment, 6 patients (75%) received corticosteroids, 6 received colchicine (75%) and 5 (62'5%) required biological drugs. Symptoms resolved completely in 4 patients (50%), 3 (37.5%) obtained partial resolution and 1 patient (12.5%) did not obtain a response.

Conclusions: Autoinflammatory syndrome is a rare entity in our environment that presents with non-specific symptoms. For this reason, it is necessary to include it in the differential diagnosis of periodic fevers, since its treatment consists of immunomodulatory drugs that modify the clinical manifestations in most of our patients.

Reference:
Sutera, D., Bustaffa, M., Papa, R., Matucci-Cerinic, C., Matarese, S., D'Orsi, C., Penco, F., Prigione, I., Palmeri, S., Bovis, F., Volpi, S., Caorsi, R., & Gattorno, M. (2022). Clinical characterization, long-term follow-up, and response to treatment of patients with syndrome of undifferentiated recurrent fever (SURF). Seminars in Arthritis and Rheumatism, 55(152024), 152024

Keywords: *undifferentiated autoinflammatory syndrome, fever, arthralgia, maculopapular rash, colchicine, corticosteroids*

[Abstract:1625]

CLINICAL PROFILES OF SYSTEMIC SCLEROSIS IN A MOROCCAN SETTING

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Introduction: Systemic sclerosis (SSc) is an orphan disease characterised by autoimmunity, fibrosis of the skin and internal organs, and vasculopathy.

Purpose: To determine the clinical, paraclinical, and therapeutic characteristics of systemic sclerosis in our population.

Methods: This retrospective study spans 11 years (2011-2021), including patients with SSc as per the 2013 ACR criteria, as well as scleroderma-overlap syndrome (SO), compiled at Mohammed VI University Hospital's Department of Internal Medicine in Marrakech.

Findings: 86 cases were included. The average age was 45.91 years with a female-to-male ratio of 13:1. All cases had cutaneous involvement, with diffuse forms (56.9%) being more common than limited forms (43.1%), of which 18.6% were classified as SO. Raynaud's phenomena occurred in 76.7% of cases. Gastrointestinal involvements were noted in 53.5%, including esophagitis and erythematous pangastritis. Pulmonary involvement was noted in 66.3%, including interstitial lung disease (50%) and pulmonary hypertension (12.8%). Renal involvement was seen in 5.8%, with 2 cases of scleroderma renal crisis. Neurological involvement was present in 12.8%, exclusively in SO. Antinuclear antibodies were detected in 83.1%, with anti-topoisomerase antibodies in 32%, and anti-centromere antibodies in 5.6%.

Patients received mainly colchicine (87.2%), corticosteroids (60.5%), and calcium channel blockers (44.1%). Cyclophosphamide was used in 39.5% for severe pulmonary involvement followed by azathioprine. A favorable or stable course was noted in 84.5% of the cases.

Conclusions: SSc is a complex autoimmune disease, making it difficult to study its multiple aspects. Further national-scale studies are necessary to better understand its profile in the Moroccan population.

Keywords: *systemic sclerosis, connective tissue diseases, autoimmune diseases*

[Abstract:1643]

DYSPNEA IN A PATIENT WITH RAYNAUD'S PHENOMENON

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A 62-year-old woman with hypertension and a history of cessation of smoking for 20 months presented dyspnea on exertion, without cough, reporting Raynaud's phenomenon in the directed history.

The echocardiogram showed asymmetric hypertrophy of the left ventricle with preserved systolic function. The chest X-ray and HRCT showed an interstitial pattern with a pattern of probable usual interstitial pneumonia (UIP). Spirometry compatible with normal except for slight impairment of diffusion (DLCO 54%). Laboratory tests: within normality. Bronchoalveolar lavage (BAL) study: 98% macrophages. Negative cytology for neoplasia. Lymphocyte populations: CD3+: 84%; CD4+: 24%; CD8+: 59%.

The study of avian and fungal serum precipitins, total and pneumoallergen-specific IgE, with normal or negative results.

Autoimmunity study showed positivity for antinuclear antibodies (ANA), at a titer of 1/320, a homogeneous nucleolar pattern with positivity for anti-Th/To antibodies (Ab).

Capillaroscopy with evidence of reduction in the number of capillaries, megacapillaries and capillary tortuosities.

In the case of this patient, the form of clinical presentation together with the results of the complementary tests performed allowed her to be classified as idiopathic interstitial pneumonia with autoimmune features (IPAF) by meeting characteristics of the clinical and serological domains.

In successive reviews, clinical manifestations were observed (such as "puffy hands", telangiectasias or vasculitic lesions in the palms) that pointed to sclerodermiform behavior in the evolutionary picture and which could be confirmed by autoimmunity (anti-Th/To antibodies) and rest of complementary tests.

Keywords: *dyspnea, Raynaud's, usual interstitial pneumonia*

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[Abstract:1660]

IDIOPATIC HEMOPTISIS

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A 70-year-old woman, with a history of HTA and DL, underwent surgery for cerebral aneurysms 40 years ago (no reports) and was admitted for massive hemoptysis with anemization.
Upon her arrival, she was started on treatment with amchafibrin and CT angiography with ground glass opacities compatible with alveolar hemorrhage without associated vascular malforation. Hemodynamically stable with new episodes of hemoptysis that are self-limiting without subsequent anemization.
A hemoptysis study was initiated, with negative thrombophilia, autoimmunity profile (ANAs, ENAs, RF, β-2 microglobulin, lupus anticoagulant, antiglomerular membrane Ac, antitransglutaminase Ac) all of which were negative in order to rule out vasculitis, renopulmonary syndrome, Lane-Hamilton among others; infectious etiology (IGRA, sputum, micobacteria and fungi negative) and echocardiogram that showed no significant alterations. FBC was requested and showed no lesions, sending samples for microbiology (fungi, β-d-Glucan and galactomannan, respiratory virus panel negative) and for cytology negative.
The patient progressed favorably with optimized treatment of levofloxacin and corticoids.
As incidents, she presented a local reaction after the fourth dose of levofloxacin. Given the doubtful unstudied history of allergy developed symmetrical lesions on the arms and axillae of erythematoedematous papuloplaques with a pseudovesicular center so topical treatment with corticosteroids was started. But given progression in the gluteal region and suspicion of toxicoderma, a skin biopsy was performed (concordant with eczema) and systemic corticosteroids were started without further progression of the lesions and gradual remission.
The patient progressed favorably without no respiratory failure or new hemoptoic episodes, maintaining a stable Hb of 11.3 and could be discharged.

Keywords: *idiopathic hemoptysis, respiratory, autoimmune*

[Abstract:1662]

THE CONSEQUENCES OF THERAPEUTIC STEPS

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A case is reported of a 93-year-old woman with HTA, DM2, diabetic nephropathy, usual interstitial pneumonia that associated chronic respiratory failure with OCD secondary to PPH of pulmonary origin, hypertensive heart disease with normal LVEF and carrier of PM due to complete BAV.
She was diagnosed of ITP in July 2022 with response to corticosteroids abd presented a relapse of the same one month later, presenting thrombopenia with an acute hematoma intracraneal asymptomatic. Corticotherapy was started again at a dose of mg/kg, with a control head CT without changes, with partial response. Because of this, it was decided to start treatment with rituximab as compassionate use with an excellent response on this occasion.
15 days after discharge, she was admitted again with significant respiratory failure as well as severe thrombopenia. Boluses of corticosteroid therapy were started again and a second dose of rituximab was administered (one day earlier than expected due to admission). From a respiratory point of view, progression of UIP plays an important role, which improves in parallel with thrombopenia with the use of corticosteroids.
Despite the initial improvement, the patient experienced a feverish spike from which blood and urine cultures, starting empirical antibiotic treatment. In less than 12 hours, isolation of Staphylococcus aureus oxaR in all blood cultures was reported. So if endocarditis is suspected in a patient with a pacemaker, antibiotic therapy is adjusted and needs echocardiogram, which there was no time to perform. Blood cultures were repeated, with SAOR persisting.
Despite everything, the patient presents a torpid evolution and poor short-term prognosis and finally died.

Keywords: *Idiopathic thrombocytopenic purpura, corticosteroid, thrombocytopenia, immunosuppression*

[Abstract:1674]

TO SOLVE A PUZZLE CASE: ANTI-EJ ANTIBODY-POSITIVE ANTISYNTHETASE SYNDROME AND RELAPSES

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The most common presentation of patients with idiopathic inflammatory myopathy is gradual onset of proximal muscle weakness associated with muscle enzyme elevations. Antisynthetase syndrome is defined by the presence of an antibody direct against one aminoacyl-transfer (tRNA) synthetases as well as by the develop of one clinical findings like an inflammatory myopathy, interstitial lung diseases or arthritis. Considered like

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overlap syndrome, it can be added mechanic's hands and Raynaud phenomenon.

We present the case of a 71-year-old man who developed recurrent cutaneous eruptions 4 years before the diagnosis. Later, he continued with onset of myalgias in both biceps associated to high serum creatine kinase levels, arthralgias in shoulders, increased of exertional dyspnea and cough. The autoimmune test verified positive for antinuclear antibody (ANA) and anti-glycyl tRNA synthetase (anti-EJ) antibodies. Before muscle histological confirmation of polymyositis, the patient started treatment with mycophenolate mofetil (MMF) diary, pulse intravenous glucocorticoids and monthly intravenous immunoglobulin (IVIG) in three consecutive days planned in outpatient care station. Due to the development of a non-specific interstitial pneumonia (NSIP) and low diffusing capacity of the lung for CO (DLCO), he kept pirfenidone as an antifibrotic agent. Despite a positive evolution and short follow-up, the patient had 2 episodes of asymptomatic hypertransaminasemia and 2 relapses of muscle weakness and respiratory distress in relation to changes in treatment and a flu episode as triggers. In acute clinical episodes, the patient received triple therapy temporarily, and subsequently MMF and oral glucocorticoids with significant clinical improvement.

Keywords: *anti synthetase syndrome, anti-EJ antibody, mechanic's hands, polymyositis, non-specific interstitial pneumonia, relapse*

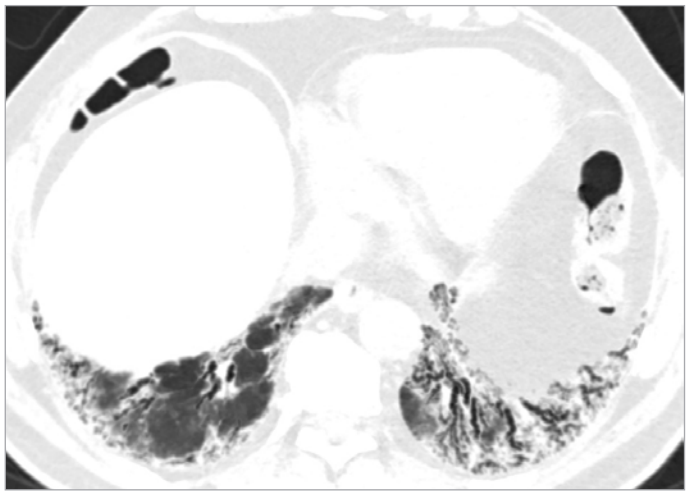


Figure 1. *Non-specific interstitial pneumonia (NSIP).*

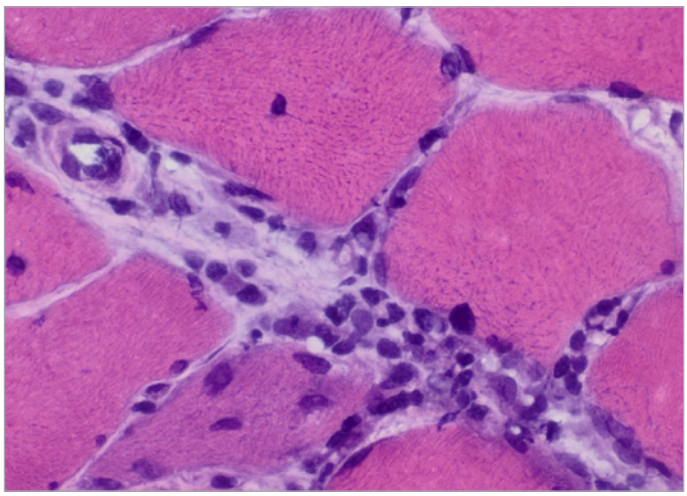


Figure 2. *Pathology of polymyositis.*

[Abstract:1690]

EPIDERMODYSPLASIA VERRUCIFORMIS REVEALING A LATE ONSET COMBINED IMMUNODEFICIENCY (LOCID)

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Introduction: we report the association of Late Onset Combined Immunodeficiency syndrome with epidermodysplasia verruciformis (EV) in a 23-year-old woman. This type of association is rare.

Case: A 23-year-old Moroccan woman, issued from a consanguineous marriage was diagnosed with Late Onset Combined Immunodeficiency (LOCID), when at age of 7 years old. She presented thousands of multiple small verrucae disseminated over her body, and continued to develop repeated respiratory and gastro-intestinal infections. A skin biopsy was consistent with epidermodysplasia verruciformis.

The blood tests showed a low absolute CD3 T cell, CD3+CD4, CD3+CD8, CD19 and CD16+CD56. The biological evolution was marked by the insurgence of a constant lymphopenia varying between 390 and 940 cells/mm^3. Hypogammaglobulinaemia was not found on two serum protein electrophoreses. In summary, of these clinical and biological data, the diagnosis of an EV secondary to a LOCID was confirmed. The patient was placed on a monthly Ig transfusion program with a good clinical evolution thereafter.

Discussion: LOCID is a type of variable common immune deficiency (VCID) that can be defined by a defect in antibody production (IgG and IgA ± IgM type), profound CD4 T-cell lymphopenia and opportunistic infections, and can be considered a distinct entity from CVID due to its particular clinical and immunological profile and its higher morbidity and mortality.

Conclusions: LOCID differs from classic VCID in its clinical and immunologic characteristics. Systematic T cell phenotype may help to discriminate such patients from those with CVID.

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Keywords: *LOCID, immune deficiency, epidermodysplasia verruciformis*



Figure 1. *Epidermodysplasia verruciformis.*

[Abstract:1694]

"CHLOROQUINE MACULOPATHY": IS THIS PHEMOMENON GIVEN ENOUGH ATTENTION?

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Chloroquine maculopathy is a not so rare complication of long-term use of chloroquine preparations in the treatment of rheumatological diseases. Its early detection is of exceptional importance for timely discontinuation of the drug in order to prevent irreversible vision damage.

The aim of this article was to present a case of a 62-year-old patient with bull's eye maculopathy as a result of long-term use of chloroquine due to systemic lupus erythematosus.

The patient presented with blurring and distortion of the pericentral vision. She was ophthalmologically evaluated with fundoscopic examination, autofluorescence, perimetry, optical coherent tomography which together with the medical history established a diagnosis of chloroquine maculopathy. Chloroquine therapy was discontinued and the patient is regularly followed by a Rheumatologist and an Ophthalmologist, with no disease progression so far.

Keywords: *chloroquine, maculopathy, systemic lupus erythematosus, Bull's eye*

[Abstract:1697]

CASE SERIES EXPERIENCE OF HEMOPHAGOCYTIC SYNDROME IN A TERTIARY HOSPITAL

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The aim of this study was a descriptive analysis of a case series of patients diagnosed with hemophagocytic syndrome (HFS) between 2018 and 2023. All those meeting diagnostic criteria for HFS at some point during their admission are extracted from the registry of patients hospitalized during that period. A total sample of 17 patients is obtained. Data on affiliation, background and prevalence of clinical and analytical manifestations were collected (see Table 1-3). To highlight some, SHF secondary to infectious disease was the most frequent in our population (64.71%), tied with that secondary to autoimmune and neoplastic disease (17.65%). Most patients had no rheumatologic (75.59%) or oncologic (58.82%) history, with non-Hodgkin's lymphoma being the most prevalent disease in our population (23.53%).Conclusions on the predictors of mortality are discussed in two other trials on this same study, carried out in our center. HFS is an entity with high mortality, as observed in our study population, and low prevalence, which explains the scarcity of literature on the subject. As a conclusion of our work, the high percentage of patients who do not receive pulsed corticosteroid therapy and the low prevalence of the use of Etoposide, despite this being one of the few first-line treatments in the management of HFS, are remarkable findings in the treatment section. It is also striking that the analytical finding with the highest incidence in our population, hypertransaminasemia, does not form part of the diagnostic criteria for SHF, according to the reference clinical practice guidelines (HLH 2004).

Keywords: *hemophagocytic syndrome, pulsed corticosteroids, hypertransaminasemia*

Variable	Sample	Saphiro-Wilk Test	Mean	Standard deviation	Mediana	Interquartile range
Age (years)	17	0.24	59.59	20.59		
Hemoglobin (g/dL)	17	0.02			8.4	1.6
Leukocytes (x10-3/mm-3)	17	0.00			1.8	16.5
Neutrophils (x10-3/mm-3)	17	0.00			1	13.1
Lymphocytes (x10-3/mm-3)	17	0.00			0.5	0.2
Platelets (x10-3/mm-3)	17	0.00			63	90
Fibrinogen	17	0.01			333	625
CRP (mg/L)	17	0.00			31	186
CRP at 7 days (mg/L)	15	0.00			13.9	44.9
CRP at 14 days (mg/L)	13	0.00				
Ferritin (ng/dL)	16	0.01			8822.5	21959
Ferritin at 7 days (ng/dL)	11	0.00			4500	17448
Ferritin at 14 days (ng/dL)	9	0.00			1070	1592
Triglycerides (mg/dL)	15	0.26	314.73	132.99		
LDH (U/L)	15	0.00			557	896
D-dimer (mcg/mL)	11	0.00			4244	5905
ESR	12	0.06	25	23.81		
Creatinine (mg/dL)	17	0.00			0.64	0.36
Total corticosteroid dose (Equivalent to mg of Methylprednisolone)	16	0.00			170.65	1697

Table 1. *Descriptive study of quantitative variables.*

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Fever	Frequency	Percentage	Cumulative Percentage
No	0	0	0
Yes	17	100	100
Adenopathies	Frequency	Percentage	Cumulative Percentage
No	6	35.29	35.29
Yes	11	64.71	100
Hepatomegaly	Frequency	Percentage	Cumulative Percentage
No	14	82.35	82.35
Yes	3	17.65	100
Splenomegaly	Frequency	Percentage	Cumulative Percentage
No	6	35.29	35.29
Yes	11	64.71	100
Arthritis/Arthralgias	Frequency	Percentage	Cumulative Percentage
No	13	76.47	76.47
Yes	4	23.53	100
Pulmonary manifestations	Frequency	Percentage	Cumulative Percentage
No	7	41.18	41.18
Yes	10	58.82	100

Table 2. Frequency table with the qualitative variables (clinical manifestations) collected in the study.

Coagulopathy	Frequency	Percentage	Cumulative Percentage
No	9	52.94	52.94
Yes	8	47.06	100
Hypertriglyceridemia	Frequency	Percentage	Cumulative Percentage
No	6	40	40
Yes	9	60	100
Hypetransaminasemia	Frequency	Percentage	Cumulative Percentage
No	1	5.88	5.88
Yes	16	94.12	100
Proteinuria	Frequency	Percentage	Cumulative Percentage
No	6	42.86	42.86
Yes	8	57.14	100
Serologies	Frequency	Percentage	Cumulative Percentage
Negative	9	52.94	52.94
EBV	2	11.76	64.71
CMV	4	23.53	88.24
Others	2	11.76	100

Table 3. Frequency Table with the qualitative variables (analytical manifestations) collected in the study.

[Abstract:1710]

GENDER DIFFERENCES IN RHEUMATOID ARTHRITIS

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Purpose: The aim of our work was to study the particularities of rheumatoid arthritis (RA) in both genders.

Methods: This was a retrospective study including patients with RA (ACR-EULAR 2010 criteria), followed in an internal medicine department between 2015 and 2022. Patients were divided into two groups: male (G1) and female (G2).

Results: Eighty-two patients were included, G1: 17 men (20.7%) and G2: 65 women (79.3%). The mean age of disease onset in G1 was 54.07±19.74 years and in G2 45.5±14.95 years. The prevalence of autoimmune diseases was higher in G2 (56.9%) than in G1 (20%); p=0.010. Rheumatoid factor (RF) was positive in similar rate in both groups. Anti-cyclic peptide antibodies (anti-CCP) were positive in 91.7% of G1 and 75.5% of G2 patients (p=0.224). Anti-nuclear antibodies (ANA) were positive in 30.8% of G1 and 54.7% of G2 patients (p=0.122). RA was erosive in 81.2% in G2 vs 75% in G1. There was no significant difference between the 2 groups in CRP, DAS28 values, RF and Anti-CCP levels. The use of different therapeutic classes was similar in the 2 groups, with no significant difference concerning corticoids, CsDMARDs and biotherapies. Extra-articular impairment was observed in approximately half of the patients in each of the two groups. There was no significant difference between the two groups concerning skin manifestations, pulmonary manifestations, serositis and scleritis.

Conclusions: A higher prevalence of autoimmune diseases and ANA positivity were associated with female patients. There was no significant biological, radiological, or therapeutic difference between the two groups.

Keywords: rheumatologic, rheumatoid polyarthritis, gender

[Abstract:1715]

CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY DURING SPONDYLARTHRITIS: IATROGENESIS OR ASSOCIATION

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Purpose: We report an observation of a chronic inflammatory demyelinating polyneuropathy (CIDP) in a patient followed for radiologic axial spondylarthritis (rAxSpA) treated with adalimumab.

Results: We report the case of a 55-year-old man followed in our service since the age of 34 for rAxSpA. He was initially on NSAIDs drugs and then on adalimumab, with clinical and biological remission. The evolution was marked by the progressive appearance of worm-type paraesthesia of the lower limbs, followed after 2 months by a proximo-distal motor deficit of the 4 limbs. Deep tendon reflexes were abolished. The electromyography had shown asymmetric bilateral sensorimotor axonal and demyelinating neuropathy of the 2 lower limbs. Lumbar puncture was not possible in front of the ankylosis of the lumbar backbone. MRI was free of anomalies. The diagnosis of CIDP was retained. An etiological investigation was carried out. The blood count was normal. No gammopathy was observed at the electrophoresis of the proteins. Tumour markers: carcinoembryonic antigen, alpha-foetoprotein, cancer antigen 19-9, and prostate specific antigen were negative. In

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addition, peripheral hypothyroidism and vitamin B12 deficiency were found. Thoraco-abdominopelvic tomography scans did not show a solid neoplasm, as did the digestive endoscopies. Anti-nuclear antibodies, anti-DNA antibodies, rheumatoid factor, anti-citrullinated antibodies, and anti-neutrophil cytoplasmic antigens were negative. The pharmacovigilance investigation concluded that the role of Adalimumab was possible.

Conclusions: In our patient, a neoplastic cause or connective tissue disease associated with CIDP was eliminated. The association with spondyloarthropathy has not been reported but remains possible. An iatrogenic cause may be responsible for demyelinating neuropathy, especially anti-TNF-alpha.

Keywords: *spondyloarthritis, neuropathy, iatrogenesis*

[Abstract:1738]

REPORT AND EXPERIENCE OF 8 CASES OF HEMOPHAGOCYTIC SYNDROME IN THE DEPARTMENT OF INTERNAL MEDICINE

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Background: A comprehensive analysis was conducted to examine cases of hemophagocytic syndrome managed within our Internal Medicine service over the last 6 years.

Methods: A retrospective observational study was undertaken, reviewing patients admitted from 2017 to 2023 and diagnosed with hemophagocytic syndrome. The study scrutinized clinical, analytical, and radiological symptoms, identified triggers, explored treatment strategies, and assessed mortality outcomes.

Results: A total of 8 cases were diagnosed, predominantly affecting women (75%) with an average age of 44.37 years. Common underlying autoimmune diseases included systemic lupus erythematosus, while infectious agents accounted for 62.5% of triggers, with notable instances of cytomegalovirus and Epstein Barr virus. Clinical features comprised fever, hepatosplenomegaly, anemia, hypertriglyceridemia, thrombocytopenia, elevated LDH, and hyperferritinemia. Hemophagocytosis was universally observed in marrow biopsies.

Conclusions: Hemophagocytic syndrome, though rare and frequently underdiagnosed, presents with diverse underlying causes. Beyond viral infections, parasitic diseases like Leishmaniasis contribute to its pathology.

Associations with hematological neoplasms and autoimmune diseases, including Still's disease and SLE, were also identified. Treatment strategies, primarily involving corticosteroid therapy, demonstrated positive outcomes in 87.5% of cases, contributing to a favorable prognosis. Recognizing the significance of early diagnosis and targeted treatment for improved patient outcomes underscores the importance of ongoing research in this uncommon pathology.

Keywords: *hemophagocytic syndrome, autoimmune diseases, Infectious triggers, clinical features, corticosteroid therapy, marrow biopsy*

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8
Age	52	30	25	60	22	45	66	55
Sex	Women	Women	Women	Women	Women	Man	Women	Man
Pathology/background	Bronchial asthma	Kikuchi disease/SLE	Still Syndrome	SLE	No one	APS/LNH lymphoblastic	No one	Autoimmune encephalitis
Immunosuppressive treatment	NO	Azathioprine	Prednisone 50 mg	Prednisone, azathioprine	NO	Azathioprine, prednisone, rituximab	NO	Lacosamide, prednisone, rituximab
Fever	YES	YES	YES	YES	YES	YES	YES	YES
Hepatosplenomegaly	YES	YES	YES	NO	YES	YES	YES	YES
Haemoglobin (g/L)	8.7	8,9	11	9,6	6,9	7,9	8,9	9,9
Platelets	99.000	330.000	69.000	63.000	110.000	17.000	201.000	99.000
LDH (UI/L)	899	439	2.091	533	327	4.200	240	904
Triglycerides (mg/dL)	202	230	286	No available	640	236	81	1.014
Ferritin (ng/mL)	1.360	1.957	33.000	4.322	2.452	1.979	261	2.338
Infectious cause	NO	Cytomegalovirus	NO	Cytomegalovirus	Epstein-Barr Virus	Leishmania	Leishmania	NO
Hemophagocytosis MO.	YES	YES	YES	YES	YES	YES	YES	YES
Treatment	Dexamethasone and Etoposide	Dexamethasone and Ganciclovir	Methylprednisolone boluses	Methylprednisolone boluses and IVIG	Dexamethasone	Methylprednisolone and IVIG	Amphotericin B and Dexamethasone	Dexamethasone, Cyclosporine, Etoposide
Exitus/Death	NO	NO	NO	NO	NO	YES	NO	NO

Table 1. *8 Cases of Hemophagocytic Syndrome in the Department of Internal Medicine.*

[Abstract:1739]

THE ROLE OF PET-IMAGING IN PATIENTS WITH SILICONE LEAKAGE FROM BREAST IMPLANTS. A CASE SERIES

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Objective: This case series explores the role of PET-imaging in patients with extensive silicone leakage from breast implants, who experience potential silicone-associated systemic symptoms.

Methods: Patients with extensive silicone leakage from breast implants and systemic health complaints underwent PET-scan imaging. Clinical assessments, including medical history, physical examination, and selective imaging studies, were conducted to exclude alternative causes for their systemic symptoms. MR imaging and/or ultrasound assessed the extent of the silicone leakage from breast implants.

Results: Fourteen patients with silicone leakage in the past, or silicone leakage from the current implants, underwent MR imaging, and/or ultrasound and PET-imaging. All presented with varied systemic symptoms, such as arthritis, Raynaud phenomenon, fatigue and muscle pains, which did not meet full diagnostic criteria for a (rheumatologic) diagnosis. Radiologic assessment identified silicone depositions in various anatomical locations, with axillary lymph nodes (n=13), intramammary (n=6), intrathoracic lymph nodes and the pectoral muscle (n=6) being the most common sites. Notably, one case exhibited silicone leakage reaching the sub- and supraclavicular, and cervical lymph nodes. In all 14 patients, PET-scan imaging revealed varied degrees of PET-

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positivity at sites corresponding to silicone depositions on MRI or ultrasound.

Conclusions: Our findings highlight the role of PET-imaging for identifying silicone depositions in patients with silicone breast implants leakage, offering a potential explanation for their unexplained systemic symptoms. The findings emphasizes the potential of PET-imaging as a valuable diagnostic tool in the assessment of silicone-related complications, contributing to enhanced clinical management and patient care.

Keywords: *silicone breast implants, PET-imaging, silicone leakage*

[Abstract:1765]

EARLY ONSET RAPIDLY PROGRESSING DEMENTIA IN A PATIENT RECENTLY DIAGNOSED WITH SJOGREN'S SYNDROME: A CASE REPORT

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Case Description and Diagnostic Pathways: We present a case of rapidly progressing dementia in a recently diagnosed Sjogren's syndrome (SJS) patient. A 65-year-old female presented to neurology outpatient clinic with symptoms including hallucinations, personality changes, urinary incontinence, which began two years ago and have significantly worsened over the last six months. Her medical history included hypertension and a spontaneous abortion. MRI results indicated diffuse cerebral and cerebellar atrophy along with mega cisterna magna. Paraneoplastic antibody and limbic encephalitis panels yielded negative results. She received a prefrontal dementia diagnosis and was referred to hematology and rheumatology due to chronically elevated ESR and CRP levels. A salivary gland biopsy revealed a classification of focus 1 consistent with SJS. Anti-Ro 52 test was highly positive, while other rheumatological tests were within normal ranges. Azathioprine 2x50 mg, methylprednisone 1x16 mg, and hydroxychloroquine 2x200 mg therapy have been started.

Discussion and Learning Points: The neurological condition was attributed to Sjogren's syndrome following the exclusion of other etiologies. In cases of rapidly progressing dementia, especially in younger patients, consideration of Sjogren's syndrome is crucial.

Keywords: *dementia, Sjogren's syndrome, prefrontal dementia*

[Abstract:1773]

RECURRENT PLEUROPERICARDITIS AFTER COVID-19 INFECTION: A CASE REPORT

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During COVID-19, some patients may develop systemic hyperinflammatory response and acute pericarditis. However, cases of recurrent pericarditis (RP) are rare.

A 47-year-old female patient was admitted with complaints of fever, chest pain that increased with breathing, and elevated acute phase reactants (CRP:173 mg/L, ESR: 72, Ferritin: 442 ng/mL, D-dimer: 2680 µg/L) for one month. Echocardiography showed pericardial effusion, a thick and fibrinous pericardium. CT showed a 1.5 cm pleural effusion on the left and involvement consistent with COVID-19 sequelae in the resolution period. Pleural effusion sampling was exudative, and cytology showed no abnormalities. There was no growth in cultures. Evaluations for tuberculosis, viral, and autoimmune serology were negative. PET-CT showed no significant uptake other than minimal FDG uptake in the pericardium (SUDmax:2.5). Anti-tuberculosis prophylaxis was started because pleural fluid was exudative, pericardial fluid contained fibrin, and her husband had a history of tuberculosis. Methylprednisolone 20 mg/day was started due to lung involvement characteristics suggestive of COVID-19 infection, considering possible COVID-19-related pleuropericardial effusion. Her complaints decreased with steroid treatment, soon to be started again when the methylprednisolone dose was reduced to 8 mg/day. Findings were compatible with recurrent pleuropericarditis secondary to prolonged inflammation after COVID-19. Anakinra 100 mg/day and colchicine tablet 1.5 mg/day were started. Clinical and laboratory complete responses were achieved and there was no recurrence with treatment in the 2nd year of follow-up.

In cases of RP developing on the background of prolonged hyperinflammatory response after COVID-19 infection, anti-IL-1 therapy is effective for treatment.

Keywords: *anti-IL-1, COVID-19, pleuropericarditis*

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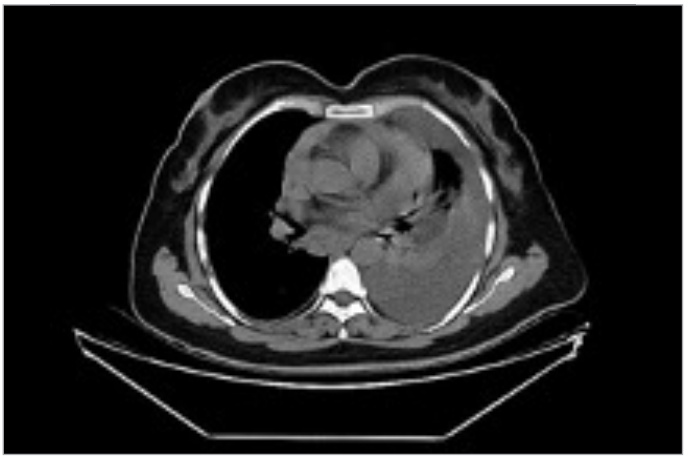


Figure 1. BT: pericardial and pleural effusion.

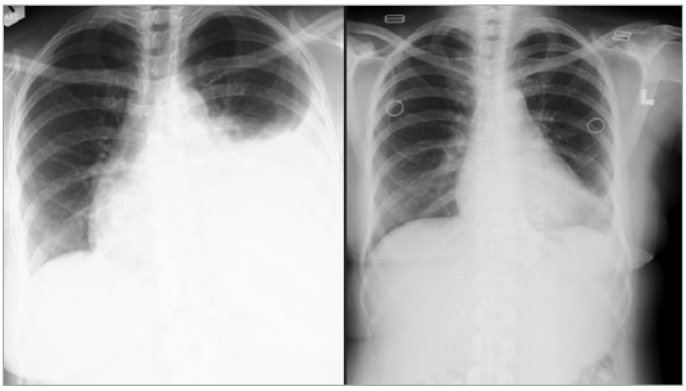


Figure 2. Chest X-ray before and after the treatment.

[Abstract:1782]

UNUSUAL SYSTEMIC LUPUS ERYTHEMATOSUS FLARE WITH ANEMIA, ARTHRITIS, ENTERITIS, NEPHRITIS AND POLYSEROSITIS

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A 25-year-old black woman without comorbidities presented herself with a year-long history of mucous diarrhea, inflammatory polyarthritis and 30 kg weight-loss. 3 months prior to admission, she was diagnosed with systemic lupus erythematosus, having started hydroxychloroquine, azathioprine and prednisolone, which she discontinued due to vomiting, 2 weeks before hospitalization. She later recurred to emergency due to symptom persistence.

Physical examination showed diminished left breath sounds and periumbilical abdominal pain. Laboratory evaluation revealed microcytic anemia, broad-base hypergammaglobulinemia, normal creatinine and hematoproteinuria (PCR 1528 mg/g). IGRA, Hepatitis B and C, EBV IgM and CMV IgM serologies were negative. C3, C4 and CH50 were low. Autoimmunity panel showed positivity for ANA (AC-1) (>1:1280), anti-dsDNA, anti-ssDNA, anti-histones, anti-Ro, anti-La and anti-RNP. Thoraco-abdomino-pelvic CT scan outlined left pleural and peritoneal effusion, diffuse mural thickening with submucosal edema of the jejunum and ileum as well as multiple iliac and retroperitoneal lymphadenopathies.

SLE flare with enteritis, nephritis and polyserositis was assumed. Methylprednisolone 1 g daily pulses were given for 3 days, followed by oral prednisolone 1 mg/kg/day and hydroxychloroquine 400 mg od. A thoracocentesis was performed with drainage of 1300 mL of pleural fluid compatible with transudate and negative for mycobacterial infection and malignant cells. The patient was transferred to the Nephrology department where renal biopsy confirmed class III lupic nephritis. Mycophenolate 500 mg bid was begun.

This complex and multi-systemic presentation (specially with lupic enteritis) represents a rare form of SLE exacerbation. We emphasize the need for a proper diagnosis and treatment to avoid relevant complications.

Keywords: *systemic lupus erythematosus, lupic enteritis, lupus flare, abdominal pain, diarrhea, vasculitis*

[Abstract:1788]

ROTATIONAL THROMBOELASTOMETRY TESTING TO EVALUATE THE HEMOSTASIS IN PATIENTS WITH SEVERE PSORIASIS

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Background: Psoriasis is an immune-mediated, inflammatory dermatosis connected to cardiovascular comorbidities. It has been repeatedly described that patients with severe psoriasis are at higher risk of thrombotic complications. Rotational thromboelastometry (ROTEM) is a viscoelastic hemostatic test that enables evaluation of in vivo hemostasis in whole blood samples. This study aimed to evaluate changes in hemostasis in psoriatic patients using ROTEM.

Methods: We performed a pilot, observational, prospective study. A total of 61 patients with severe psoriasis (PASI>15) were enrolled in this study. The control group consisted of 60 healthy blood donors. Blood samples were tested with ROTEM Gamma (Pentapharm GmbH, Munich, Germany) analyzer using INTEM, EXTEM, and FIBTEM reagents. We measured clotting time (CT), clot-forming time (CFT), and maximum clot firmness (MCF).

Results: The group of psoriatic patients involved 30 men and 31

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women with an average age of 51 (ranging from 19 to 85). We compared patients with severe psoriasis to healthy controls. Our analysis revealed significant changes in the following parameters between healthy controls and psoriatic patients: INTEM-CT (199 ± 66.1 vs. 183 ± 26.6, p<0.001), INTEM-CFT (84.9 ± 26 vs. 74.5 ± 17.3, p<0.001), EXTEM-CT (71 ± 10.8 vs. 74 ± 10.2, p<0.05), EXTEM-MCF (63 ± 4.42 vs. 66 ± 5.44, p<0.05), FIBTEM-CT (62 ± 9.28 vs 67 ± 7.68, p<0.05), and FIBTEM-MCF (16 ± 3.82 vs 17 ± 5.45, p<0.05) when compared to healthy controls.

Conclusions: We observed abnormal changes in hemostasis using rotational thromboelastometry in patients with severe psoriasis.

Keywords: *severe psoriasis, rotational thromboelastometry, venous thromboembolism*

[Abstract:1791]

A PUZZLE OF A FEBRILE LADY

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Case Description: 66-year-old lady with 2-year history of monthly cyclical fevers and joint pain was admitted with non-specific abdominal pain, diarrhoea and breathlessness with supplementary oxygen requirement. Inflammatory markers were raised including ferritin around 20,000. She had history of SLE & Sjogren's but her hydroxychloroquine (HCQ) was recently stopped as she developed Stevens-Johnson syndrome as a reaction.

Clinical Hypothesis: For possible immunosuppression due to HCQ, intra-abdominal sepsis was first considered with wide differentials of PE, haemato-oncological & rheumatological diseases.

Diagnostic Pathways: Blood cultures and infection panels including viral, sexual and TB screen were sent. Gram-negative bacterial growth was identified & treated. Despite antibiotics, she had ongoing temperature spikes. CT of abdomen and pelvis showed bilateral pleural effusions, splenomegaly and enlarged lymph nodes. CTPA ruled out PE but showed bilateral plural effusion and multiple lymph nodes with splenomegaly. TTE was unremarkable.

PET scan showed metabolic activities in multiple lymph nodes but no evidence of malignancy. For concern of hemophagocytic lymphohistiocytosis (HLH) or lymphoma, haematology was involved. H-score showed low probability of HLH. Lymph node biopsy only showed reactive changes. Bone marrow biopsy did not show obvious macrophage activity.

Adult-onset Still's disease (AOSD) was provisionally diagnosed and steroids were trialled by rheumatology but there was no response. After MDT discussion, anakinra was started with good clinical and biochemical response.

Discussion and Learning Points: With a systematic stepwise approach, infections, malignancies and inflammatory conditions were investigated in this complex presentation. MDT including histopathologists helped with diagnosis. Biologics use appropriately controlled this rare condition of AOSD.

Keywords: *fever, adult-onset Still's disease (AOSD), rheumatological, haematological, infections*

[Abstract:1798]

TEMPORAL ARTERITIS WITH CNS VASCULITIS PRESENTING AS DIPLOPIA AND PTOSIS: A RARE CASE REPORT

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A 62 year-old woman with hyperlipidemia presented herself with a 4-week tale of a novel temporal headache, scalp tenderness, jaw claudication, sporadic fever and a day-long binocular diplopia. Upon physical scrutiny, her superficial temporal arteries proved tender. The right eye suffered dextro and supraversion resulting in oblique diplopia and ptosis of the right eyelid. The remainder exam was unremarkable. Laboratory assessments unveiled leukocytosis, ESR 48 mm/h and CRP 15.99 mg/dL. Cranial and cervical CT scans with angiography showed no abnormalities.

Promptly, a methylprednisolone pulse was administered, succeeded by oral prednisolone 40 mg od. Over the ensuing 2 weeks, comprehensive workup was undertaken. Blood cultures were negative, and CSF analysis, including Herpesviridae, microbiological culture, and oligoclonal bands, demonstrated normalcy. Negative results were obtained from ANA, ANCA, IGRA, HBV, HCV, and HIV tests. Cranial MRI demonstrated hyperintense punctiform foci (T2 FLAIR), exhibiting bi-hemispheric, fronto-parietal topography congruent with vasculitis. A temporal artery biopsy, conducted within two weeks under corticosteroids, corroborated fragmentation of the internal elastic lamina and chronic inflammation. Giant cells were absent. The diagnosis of temporal arteritis was confirmed. Four months into follow-up and presently undergoing a 52-week corticosteroid tapering regimen, the patient has attained full clinical remission, encompassing resolution of the right third cranial nerve palsy and the normalization of inflammatory markers (VS 1 mm/h). Though uncommon, diplopia may manifest in 1-19% of temporal arteritis patients, with the occurrence of oculomotor palsy, especially coupled with ptosis, being a rarity. It must be faced as an ophthalmological emergency, instigating expedient corticosteroid therapy.

Keywords: *temporal arteritis, giant cell arteritis, central nervous system vasculitis, diplopia, cranial nerve palsy, ophthalmological emergencies*

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[Abstract:1803]

FEVER, ARTHRALGIA AND RASH IN ADOLESCENT WOMEN

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A 16-year-old woman, with no personal history of interest, who came to the emergency room with fever along with arthralgia, weakness in the lower limbs, and odynophagia lasting 48 hours. Likewise, she has spontaneous ecchymosis on the trunk and limbs and bilateral eyelid edema.

On examination, there was bilateral palpebral edema with a violet tint, a hyperemic pharynx without exudates, normal cardiopulmonary auscultation, hematoma in 2-3 left intercostal space, left hip and scapula, inability to stand due to pain in the lower limbs, without arthritis, with strength, sensitivity and reflexes preserved.

The analysis highlights only RCP 77.8 mg/L, CK 200U/L, aldolase 8.2 U/l and Anti-streptolysin O 669 U/ml.

For all this, she was admitted to the hospital for study.

During her hospital stay, she had skin lesions, low-grade fever that improved with metamizole, and generalized arthralgias. Microbiological study is requested (pharyngeal exudate, blood cultures, PCR of mycobacteria in bronchial aspirate, serologies, IGRA) with negative results, as well as a negative autoimmunity study for ANA, ENAs, antibodies to dermatomyositis, scleroderma, related to overlap syndromes and ANCA.

The skin biopsy shows acute vasculitis of a medium-caliber vessel with the absence of immunoglobulin and complement deposits, suggestive of panarteritis nodosa (PAN). Treatment was started with boluses of methylprednisolone and subsequently prednisone in a descending regimen associated with methotrexate.

Currently, the patient has presented a significant improvement in symptoms after starting treatment, maintaining prednisone on a descending regimen, associated with methotrexate.

Keywords: *vasculitis, polyarteritis nodosa, autoimmune diseases*

[Abstract:1811]

ENDOVASCULAR TREATMENT OF AN ILIAC ARTERY ANEURYSM IN A RHEUMATOID ARTHRITIS PATIENT

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Introduction: Rheumatoid arthritis (RA) patients have increased risk of cardiovascular mortality. Here we present an endovascular treatment of an iliac artery aneurysm in a RA patient.

Case Report: A 69-year-old female patient presented with abdominal pain which was apparent on lower quadrants. She was diagnosed with hypertension, diastolic heart failure and seropositive RA. Her treatment consisted in perindopril, amlodipine, acetylsalicylic-acid and clopidogrel. No anti-inflammatory agent was used considering on remission for RA. On physical examination, the patient had bilateral ulnar deviation of the metacarpopharyngeal joints but no signs of arthritis, there was no abdominal tenderness or ascites.

Abdominal ultrasonography showed multiple gallstones and grade one hepatosteatosis. Computer tomography angiography showed an aneurysm in the right internal iliac artery with a diameter of five centimeters leading to 70% stenosis in the artery. We claimed the aneurysm is the cause of abdominal pain and considering age and comorbidities of the patient a minimally invasive option, coil embolization, was preferred for the treatment.

On the diagnostic digital subtracted angiography both iliac arteries were atherosclerotic but patent and right iliac artery showed an aneurysmatic dilatation and irregularities. Arterial embolization was performed, and control imaging studies showed reduced blood flow through aneurysmal sac. After the procedure no complications were seen, and the patient's presenting complaint was resolved as well.

Results: RA can cause cardiovascular complications and accelerated atherosclerosis even in remission state. Especially in the elderly patients with multiple comorbidities, endovascular treatments can lead to reduction in the post-operative complications of surgery.

Keywords: *rheumatoid arthritis, endovascular treatment, arterial aneurysm*

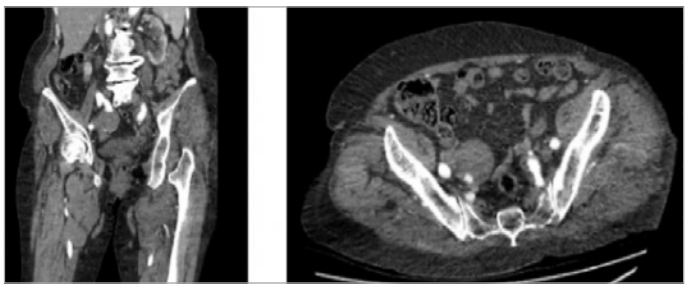


Figure 1. *Iliac aneurysm before the procedure.*

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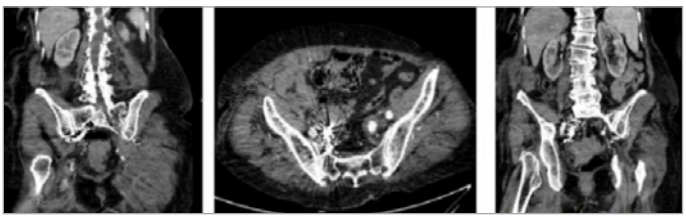


Figure 2. Iliac aneurysm after the coil embolization.

[Abstract:1815]

DECREASE IN HEMOGLOBIN LEVELS IN ACUTE IDIOPATHIC PERICARDITIS: A MODEL OF ANEMIA OF ACUTE DISEASES?

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Background: Anemia of chronic diseases (ACD) is a common condition. Anemia of acute diseases has not been thoroughly described in literature. In our clinic, we observed some patients with a decrease of hemoglobin during acute attack of pericarditis, followed by a return to basal values in few days.

Materials and Methods: We consecutively enrolled patients with idiopathic pericarditis followed at Fatebenefratelli Hospital and Papa Giovanni XXIII Hospital from January 2010 to April 2023, and retrospectively analyzed data. As primary endpoint, we analyzed Hb difference between acute attack and remission, while, as secondary endpoints, we correlated this variation with indices of inflammation and serosal involvements, using uni- and multivariate analysis.

Results: We enrolled 62 patients (median age: 39 years). Hb levels during attack were lower than those during remission [p<0.001], and were associated with an elevation of CRP [p=0.002], neutrophilia [p<0.001], instead the reduction of Hb was not correlated with presence of pericardial [p=0.846], pleural [p=0.145] and peritoneal [p=0.629] involvements. In multivariate analysis, only CRP elevation was associated with variation of Hb [β=0.336, p=0.007].

Discussion: Inflammation during acute attack of pericarditis was associated with transient anemia and the decrease of Hb levels significantly correlates with neutrophilia and CRP values. The reason for this could be related to the high IL-1 production by the inflammasome, which is hyperactivated in the context of acute pericarditis.

Conclusions: Our study showed that Hb values decrease during acute attack of pericarditis, in this way acute pericarditis may represent a pathogenetic model of anemia of acute diseases.

Keywords: *idiopathic pericarditis, hemoglobin, anemia*

Demographic characteristics	
Age at event	39 (26-60)
Gender F:M - n (%)	30 (48.4%): 32 (51.6%)
Smoking habits - n (%)	27 (43.6%)
Comorbidities - n (%)	22 (35.5%)
Clinical characteristics	
Pericardial effusion - n (%)	48 (77.4%)
Size of pericardial effusion at index attack [mm] - median (IQR)	6 (3.75 - 10.75)
Cardiac tamponade at the index attack - n (%)	2 (3.2%)
Pericardiocentesis at the index attack - n (%)	4 (6.5%)
Myocardial involvement - n (%)	12 (19.4%)
Pleural involvement - n (%)	28 (45.2%)
Peritoneal involvement - n (%)	8 (12.9%)
Laboratory findings	
Antinuclear antibody positivity, titer >1:160 - n (%)	7 (11.3%)
Maximum CRP [mg/L] - median (IQR)	
Index attack	70 (29-197.25)
Remission	5 (2-5)
Difference (Delta CRP)	68 (24-174.75)
White blood cells count (cells/mm3) - median (IQR)	
Index attack	11400 (6660-13225)
Remission	7170 (6275-8030)
Difference (Delta leukocytes)	4359 (1673-6187)
Absolute neutrophil count [cells/mm3] - median (IQR)	
Index attack	8694 (5826-10708)
Remission	4150 (3616-5993)
Difference (Delta neutrophils)	4390 (1410-6717)
Absolute lymphocytic count [cells/mm3] at index attack - median (IQR)	1640 (1210-3067)
Neutrophil-lymphocyte ratio, NLR, at index attack-median	5.93 (3.27-7.41)

Table 1. Demographic, clinical and laboratory findings of the study population.

Hemoglobin levels	Hb [g/dl] - median (IQR)	p-value*
Index attack	12.0 (11.2 to 13.4)	
Remission	13.7 (13.1 to 14)	
Difference (Delta Hb)	1.4 (0.9 to 1.98)	<0.001

Table 2. Variations in hemoglobin levels (Delta Hb).

Clinical features	Delta Hb during attack [g/dl] - median (IQR)	Delta Hb in remission [g/dl] - median (IQR)	p-value
Pericardial effusion	1.38 (0.97 to 2.06)	1.75 (0.45 to 1.94)	0.846
Pleural involvement	1.68 (1.02 to 2.56)	1.35 (0.90 to 1.80)	0.145
Peritoneal involvement	1.65 (0.77 to 2.39)	1.40 (0.94 to 1.90)	0.629
Antinuclear Ab positivity >1:160	1.20 (0.90 to 3.20)	1.40 (1.00 to 1.95)	0.879

Table 3. Correlation between hemoglobin and clinical features of pericarditis.

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Correlations of Delta Hb with	ρ (95% IC)	p-value
Age at event	0.005 (-0.253 to 0.261)	0.972
Delta CRP	0.391 (0.149 to 0.589)	0.002
Delta white blood cells	0.304 (0.051 to 0.520)	0.016
Delta neutrophils	0.426 (0.190 to 0.615)	<0.001
Maximum CRP at index attack	0.387 (0.145 to 0.586)	0.002
Maximum white blood cells count at index attack	0.279 (0.023 to 0.500)	0.028
Maximum neutrophil count at index attack	0.374 (0.130 to 0.576)	0.003
Maximum lymphocytic count at index attack	-0.064 (-0.315 to 0.196)	0.624
NLR at index attack	0.278 (0.022 to 0.499)	0.029
Size of pericardial effusion at index attack	0.108 (-0.153 to 0.355)	0.404

Table 4. *Correlation of variations in hemoglobin levels with other laboratory variables.*

[Abstract:1826]

COMPARATIVE ANALYSIS OF A CASE SERIES OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS DUE TO INFECTIOUS AND AUTOIMMUNE ETIOLOGY: SEARCH FOR PROGNOSTIC FACTORS

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The aim of this study was to analyse variables with possible prognostic implications in patients diagnosed with haemophagocytic syndrome (HLH) in the last 5 years in a tertiary hospital. An observational, retrospective and analytical study of 14 patients was carried out, dividing the groups into HLH associated with infections (I-HLH) and systemic diseases (A-HLH). The outcome, defined as partial remission or relapse, was compared between the two groups.

Results indicated that age and gender were not significantly associated with outcome in either group. Hyperferritinaemia, elevated C-reactive protein and hypertransaminemia showed a strong correlation with outcome in both groups. In the case of anaemia, correlation with relapse was only found in the A-HLH group, as was elevated lactate dehydrogenase enzyme (LDH) and erythrocyte sedimentation rate (ESR).

HLH is a low incidence, high mortality entity characterised by immune overstimulation with multiple associative causes, with viral infections, especially Epstein-Barr virus, being the most common trigger. Systemic diseases constitute another important etiological group, the two entities with the strongest association are systemic lupus erythematosus and adult Still's disease, acting as predisposing factors for immune dysregulation. Evidence on prognostic factors is limited. Age, hyperferritinemia, underlying malignancy, delay in establishing treatment and clinical-analytical signs of shock at diagnosis are poor prognostic factors, as we corroborated in our series. The study by Arca. M et al, adds hypertransaminemia and LDH elevation as predictors, also found in our series. The low suspicion and mortality related to HLH underline the need to search for prognostic factors as a priority.

Keywords: *haemophagocytic lymphohistiocytosis, haemophagocytic syndrome, macrophage activation syndrome, HLH, prognosis*

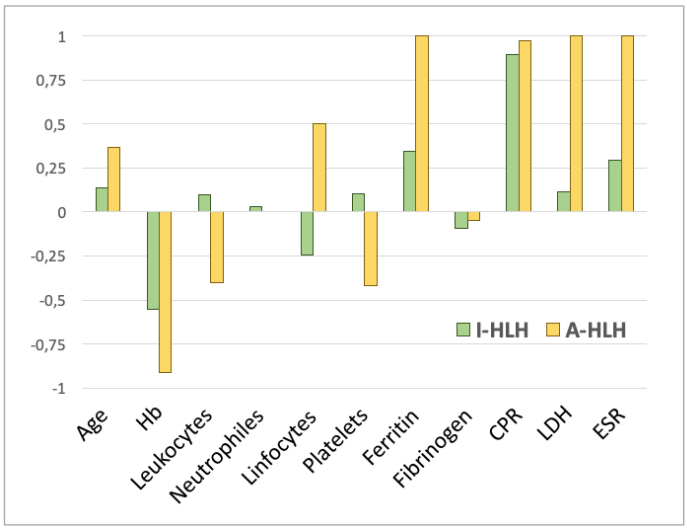


Figure 1. *Graphical representation of comparative analysis between I-HLH and A-HLH segmented according to outcome (relapse or partial remission) using Pearson's linear coefficient.*

[Abstract:1837]

A SERIOUS COMPLICATION OF RHEUMATOID ARTHRITIS: ATLANTOAXIAL SUBLUXATION

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Aim: We wanted to draw attention to a complication of RA that may have serious consequences if not considered.

Case Description and Diagnostic Pathways: A 68-year-old female patient was referred to our outpatient clinic for preoperative evaluation for a cholecystectomy operation. It was learned that she had rheumatoid arthritis (RA) for fifteen years and was not taking her medication regularly. On examination, she had erosive deformities in her hands. The patient who also had neck pain was found to have atlantoaxial subluxation (AAS) on the preoperative vertebral X-ray (Figure 1). The patient was referred to neurosurgery department and RA treatment was also planned.

RA is a chronic systemic inflammatory disease primarily affecting bones, synovial joints, and ligaments. Unlike spondyloarthropathies, rheumatoid arthritis does not involve the axial skeleton except the atlantoaxial joint. Atlantoaxial subluxation (AAS) is an important and potentially life-threatening complication of erosive RA. Instability in the atlantoaxial joint can result in multiple neurologic symptoms, including quadriparesis or quadriplegia. It is defined when the gap between the odontoid

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process and the atlas arch from C2 is more than 3 mm on a hyperflexion and hyperextension cervical lateral radiograph. The radiograph of another patient is shown in figure 2.

Discussion and Learning Points: In patients with rheumatoid arthritis, neck pain should be questioned in the preoperative evaluation and if neck pain is present, neck radiography should be performed in terms of AAS.

Keywords: *atlantoaxial, subluxation, rheumatoid arthritis*



Figure 1. *Lateral X-ray of the cervical spine in hyperflexion showing a C1-C2 diastasis.*



Figure 2. *Lateral X-ray of the cervical spine in hyperextension*

[Abstract:1840]

COEXISTENCE OF ANTI-TOPOISOMERASE I AND ANTI-CENTROMERE ANTIBODIES IN A PATIENT WITH DIFFUSE SYSTEMIC SCLEROSIS

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Introduction: Anti-topoisomerase I (ATA) and anti-centromere (ACA) antibodies are markers of systemic sclerosis (SSc). They are reported to be mutually exclusive. We report an infrequent case of coexistence of ATA and ACA in a patient with diffuse SSc.

Case Report: A woman aged 17 years consulted for muscle weakness. Diagnosis of dermatomyositis was suspected in front of abnormal findings at electromyography and histological feature of the muscle biopsy. Creatinine phosphokinase was in the normal range but sedimentation rate was increased. Antinuclear antibodies (ANA) were positive with a titer of 1/1280 and a speckled pattern. ANA typing showed the following specificities: ACA(+++), ATA(+++), anti-Sm(+++), anti-Sm/RNP(+++), anti-SSA(+++) and anti-Ro52(+++). Dot-myositis was negative apart from a positivity of anti-Ro52(+++) and a weak positivity of anti-Ku(+).

Diagnosis of dermatomyositis was not retained. A SSc was suspected in front of puffy fingers, fingertip lesions, articular involvement but there were no cutaneous sclerosis nor Reynaud's phenomenon nor telangiectasia.

Three years after that, the patient was hospitalized for worsened trophic disorders, diffuse cutaneous sclerosis, Reynaud's phenomenon and abnormalities at capillaroscopy. ANA typing showed the previous specificities and identified also anti-native DNA, anti-nucleosomes and anti-histones antibodies. The diagnosis of diffuse SSc was made. The patient did not have renal, cardiac or neurological complications. Pulmonary and digestive involvement was probable. An overlap syndrome (SSc/another connective tissue disease) was evoked but not confirmed.

Conclusions: Generally, ATA and ACA are associated with diffuse and limited forms of SSc, respectively. Only few cases of coexistence of these two autoantibodies are reported in the literature.

Keywords: *anti-centromere, anti-topoisomerase I, systemic sclerosis*

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[Abstract:1846]

URTICARIA: A RARE CUTANEOUS FINDING OF FAMILIAL MEDITERRANEAN FEVER

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Case Description: A 26-year-old male patient with no records of previous disease referred to us with rashes on his body. His medical history revealed he had complaints of intermittent abdominal pain and fever lasting 2-3 days for 7 years accompanied by a history of Familial Mediterranean Fever(FMF) in his mother, uncle and sister. The physical examination of the patient, other system examinations were normal except for the rash.

Clinical Hypothesis: A renal amyloidosis case presented with urticaria.

Diagnostic Pathways: At the time of admission, his complete blood count was normal and his complete urinalysis was positive for protein. CRP: 129 mg/L, albumin: 22 mg/dL, LDL: 215 mg/dL, and other biochemistry results were normal. In 24-h urine collection 8.1 grams of proteinuria was detected and urinary ultrasonography was performed to see that the kidney sizes were normal. Furthermore, *MEFV* gene mutation analysis was sent from the patient and a kidney biopsy was planned with the preliminary diagnosis of renal amyloidosis. MEFV gene mutation analysis is M694V resulted homozygous. Kidney biopsy showed 19 glomeruli, 6 of which were glomerular sclerotic, Congo negative. In immunohistochemical examination, strong Amyloid A was reported as amyloidosis. The patient's toxic appearance and existing complaints improved with colchicine and IL-1 antagonists.

Discussion and Learning Points: Urticaria is the least common cutaneous finding in attacks of FMF and should be considered in the differential diagnosis of autoinflammatory diseases. Based on medical history and laboratory examinations, FMF was considered in this patient, and a definitive diagnosis was made with renal biopsy.

Keywords: *amyloidosis, familial Mediterraen fever, urticaria*



Figure 1. *The patient presented with localized, irregularly bordered, raised urticarial lesions on the back at the time of admission.*

[Abstract:1859]

RENAL INVOLVEMENT IN PRIMARY SJOGREN'S SYNDROME: THE UNDERESTIMATED COMPLICATION

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Summary: Primary Sjogren's syndrome (pSS) is an autoimmune disorder that results in exocrine gland dysfunction. Renal disease is thought to be a rare extraglandular manifestation, but it is likely to be underdiagnosed due to its paucisymptomatic presentation. Tubulointersticial nephritis (TIN) and glomerulonephritis are the most common entities.

Purpose: To determine the prevalence of renal involvement in pSS and to describe the baseline characteristics of patients with pSS-related renal disease.

Methods: A retrospective analysis of clinical and immunologic characteristics of pSS patients from 1993 to 2023 in our centre was performed. 252 patients were included. Overlap with other autoimmune diseases was excluded.

Findings: Renal disease was identified in 22.2% of our sample. Median age and serum creatinine at diagnosis were 65.6 and 3.58mg/dl respectively, and 90% were women. Most had no proteinuria, hematuria o hydroelectrolyte abnormalities. TIN was the main manifestation (92.9%), followed by glomerulonephritis

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(5.36%). However, only two patients underwent renal biopsy. Articular involvement was the most frequent extraglandular manifestation (53.6%). Eighty-two percent of patients had antiRo, 61% rheumatoid facto, 58,9% anti-La/SSB, 50% cytopenias and 42% hypergammaglobulinaemia. Twenty-one patients received immunosuppressive treatment, 25.95% presented with worsening renal function, 43% had thrombotic or infectious complications and nine died during follow-up.

Conclusions: This study shows that renal disease may be more prevalent in pSS than previously thought. It is associated with the expression of immunologic markers of lymphocyte B activation. Finally, it highlights the importance of actively screening for renal involvement in order to prevent its occurrence and the associated morbimortality.

Keywords: *Sjogren, renal disease, prevalence*

[Abstract:1861]

PREDICTIVE FACTORS OF RENAL DISEASE IN PRIMARY SJOGREN SYNDROME

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Summary: Renal disease in primary Sjogren's syndrome (pSS) is associated with higher rates of chronic kidney disease (CKD), hospitalisation and mortality. Studies have attempted to identify factors that predict renal involvement in pSS, but their results are conflicting and only anti-LA/SSB was associated with renal disease in a recent meta-analysis.

Purpose: To identify factors that could predict the development of renal disease in pSS.

Methods: A retrospective analysis of pSS patients diagnosed between 1993 and 2023 at our centre was conducted. A univariate study and a multivariate analysis using a multiple logistic regression model were performed.

Findings: Results showed that 56 out of 252 patients had renal involvement.

Both in the univariate analysis and in the logistic regression model, lymphadenopathy (OR 3.45, 95% CI 1.22-9.55, p=0.017), lung disease (OR 4.41, 95% CI 1.40-13.87, p=0.010), peripheral nervous system involvement (OR 6.50, 95% CI 1.11-34.13, p=0.028) and rheumatoid factor (OR 3.11, 95% CI 1.31-7.73, p=0.012), were independently associated with renal disease. In the multivariate analysis, the variables antiLa/SSB (OR 1.84, 95% CI 0.73-4.83, p=0.204) and cryoglobulinemia (OR 2.97, 95% CI 0.57-16.26, p=0.196) did not reach statistical significance, despite being significant in the univariate analysis and belonging to the logistic regression model, possibly due to a limitation of the sample size.

Conclusions: Our study suggests that the aforementioned extraglandular manifestations may be potential predictive factors of renal involvement in pSS.

Keywords: *Sjogren, renal disease, predictive factors*

[Abstract:1904]

POLYARTERITIS NODOSA: A CASE REPORT

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A 59-year-old man presented with anorexia and weight loss, arthralgias, tinnitus and hypersensitivity of the scalp, as well as sudden testicular pain. He began with blurred vision in the right eye and later in the left eye, and was assessed by the ophthalmologist, who diagnosed bilateral exudative retinal detachment. During admission, the patient presented elevated blood pressure (180/100 mmHg) associated with confusion symptoms. No personal history of interest. Physical and neurological examination with no alterations. Laboratory tests included CRP 99 mg/dl, ferritin 406 ng/dl, normal tumour markers, normal proteins and complement, subnephrotic proteinuria of 0.5 g/24h, negative autoimmunity profile and serology. Additional tests included a lumbar puncture showing an absence of cells and hyperproteinorrachia. The body CT scan showed renal infarcts with no other findings and MRI of the skull showed findings compatible with acute ischaemic lacunar lesions. Based on the above findings, the clinical suspicion of polyarteritis nodosa was reached.

PAN is a systemic necrotising vasculitis usually affecting medium-sized arteries, not associated with ANCA antibodies. It is a rare entity, with an annual incidence of 2-9/1,000,000 of the population. It mainly affects adults aged 40-60 years, with a peak incidence in the sixth decade of life. It is predominantly male and is also associated with genetic disorders such as ADA2 deficiency and VEXAS syndrome. Treatment depends on the severity of involvement, the therapeutic arsenal being corticosteroids, immunosuppressants such as cyclophosphamide and recently the use of rituximab and other monoclonal antibodies has been incorporated. Mortality is not negligible, up to 25%.

Keywords: *vasculitis, testicular pain, exudative detachment of the retina*

 DOI: 10.12890/2024_V11Sup1

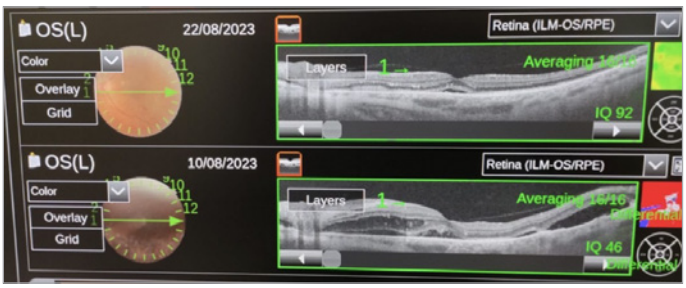


Figure 1. OCT left eye before and after the treatment.

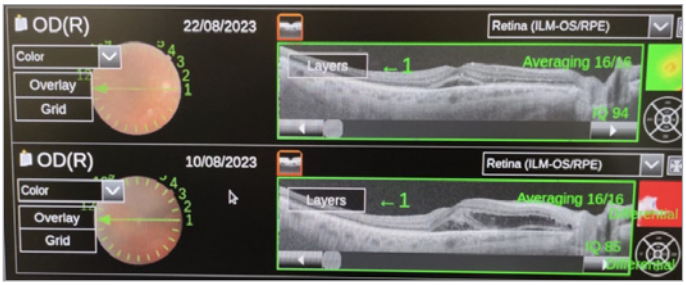


Figure 2. OCT right eye before and after the treatment.

[Abstract:1906]

URTICARIAL VASCULITIS AT THE ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS: SPECIFIC THERAPEUTIC CONSIDERATIONS

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A 21-year-old Caucasian woman, with a history of polycystic ovary syndrome (POS) and subclinical hypothyroidism, consulted for skin rash, polyarthralgia, joint stiffness and dactylitis, along with self-limited episodes of eyelid angioedema and Raynaud's syndrome since 2020.

Physical examination revealed swelling and pain in the carpus, proximal interphalangeal joints, elbows, shoulders and both knees along with dactylitis.

Analytically, she presented consumption of C3, ANA 1/1280 (coarse granular pattern) and anti-RNP-U1 antibody. Remaining tests were normal.

At this stage, it was considered a case of debut of systemic lupus erythematosus (SLE) as it had 15 points according to EULAR/ACR 2019 classifying criteria and treatment with hydroxychloroquine and glucocorticoids was started, with poor evolution. Treatment was escalated with azathioprine and tacrolimus at variable doses, although joint symptoms could not be controlled. So treatment with belimumab 400 mg/week/sc was started, with improvement in the joint symptoms.

In the subsequent months, the patient developed dyspnea and respiratory difficulty that were attributed to treatment with belimumab, which was discontinued. Since then, the skin lesions worsened and the disabling joint pain reappeared. The skin biopsy showed that it was hypocomplementemic vasculitis urticaria (low C1q) and allergy to belimumab was ruled out. After reintroducing the drug, the lesions disappeared and the patient improved overall. Belimumab is a human monoclonal antibody with indication for systemic lupus erythematosus and lupus nephritis in the latest EULAR guidelines with the same level of recommendation as the traditional treatments and that, in our experience, has been useful in the treatment of urticarial vasculitis.

Keywords: *urticarial vasculitis, systemic lupus erythematosus, autoimmune diseases*



Figure 1. Maculopapular lesions suggestive of urticarial vasculitis.



Figure 2. Maculopapular lesions suggestive of urticarial vasculitis.

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[Abstract:1912]

DYSPNEA ON EXERTION AND COUGH, NOT EVERYTHING IS SARS-COV2 INFECTION

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We report the case of a 52-year-old woman under follow-up by the pulmonology department for residual cough and dyspnea after second SARS-CoV-2 infection and evaluated by Internal Medicine for dry eye syndrome with xerostomia and weight loss of 36 kg in 8 months. Body-CT was requested, with a finding of bronchi of caliber at the high limit of normal and discreet. Respiratory functional tests were performed, which showed objective diffuse and extensive involvement of the small airway, bronchioles, and mixed spirometric pattern with severe obstruction, FEV1/FVC: 52.90%, with negative bronchodilator response, along with alteration in diffusion, DLCo/VA: 48.9%. In fiberoptic bronchoscopy, mucopurulent discharge and inflammatory mucosa and biopsies were taken. Physical examination revealed tachypnea with speech and cough along with pulmonary auscultation with widespread hypophonesis. Additionally, he had a dropped left foot with hypoesthesia and decreased strength in the extension. The patient entered the Internal Medicine unit for further tests. Ventilation/Perfusion Scintigraphy requested to rule out microthrombi secondary to SARS-CoV-2 infection was compatible with significant alteration of pulmonary ventilation, suggestive of severe airflow obstruction, without findings indicative of pulmonary thromboembolism. The electroneuromyogram described a lesion axonal and partial motor fibers of the left peroneal nerve compatible with incipient mononeuritis multiplex. During admission, the result of the pathological anatomy of the bronchial mucosa with infiltrate was received: chronic inflammatory. The autoimmunity test revealed the presence of antibodies anti-muscarinic cholinergic M3 receptor.

The patient was diagnosed with "bronchiolitis obliterans and mononeuritis multiplex of the peroneal nerve left, secondary to Sjogren's syndrome." She received treatment with methylprednisolone boluses intravenous, followed by rituximab.

Keywords: *Sjogren's Syndrome, bronchiolitis obliterans, mononeuritis multiplex*

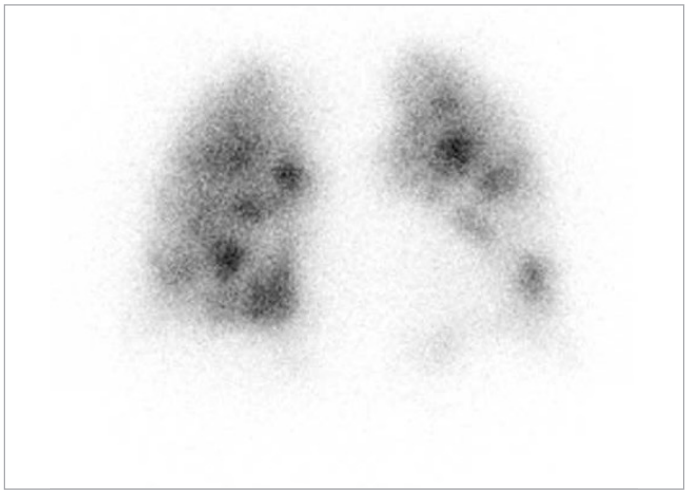


Figure 1. *Section of lung scintigraphy, antero-posterior view, showing serious alterations ventilation.*

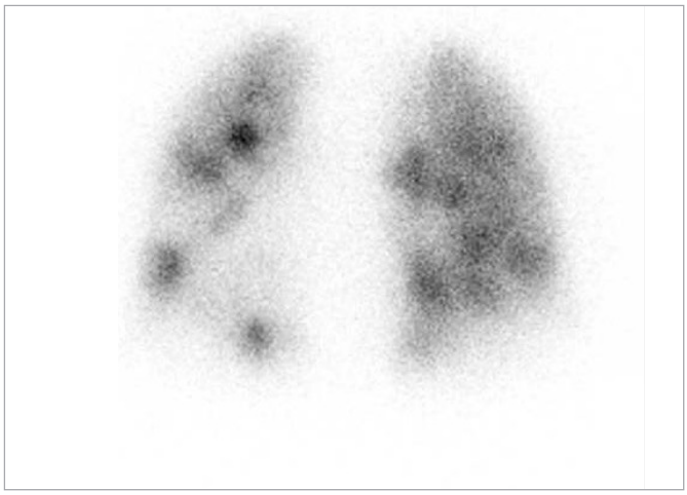


Figure 2. *Section of lung scintigraphy, posteroanterior view, showing serious alteration is observed ventilation.*

[Abstract:1931]

SYSTEMIC LUPUS ERYTHEMATOSUS AND STEEL HEART

Ana María Aldea Gamarra, Marta Salas Sánchez, Luis Antonio Álvarez Sala Walter, María Victoria Villalba García, Cristina Ausín García

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Case Description: We report the case of a 73-year-old woman with a history of systemic lupus erythematosus (SLE) with triple positivity for antiphospholipid autoantibodies consults for a 3-month history of asthenia, progressive dyspnea, and fever-like sensation. A complete study was conducted, including a microbiological and autoimmune screening. Blood tests showed significant increase of biochemical markers of myocardial injury, with no significant elevation of inflammatory markers.

Chest X-ray resulted normal, while echocardiogram discovered a slightly decreased left ventricular ejection (51%). Holter monitor registered very high density ventricular extrasystole.

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Cardiovascular magnetic resonance imaging was inconclusive, showing images compatible with a possible arrhythmogenic cardiomyopathy without being able to exclude extensive myocarditis.

Clinical Hypothesis: Given the clinical findings, we considered lupus myocarditis versus coronary heart disease.

Diagnostic Pathways: A computed tomography and positron emission tomography (PET/CT) was performed, showing no cardiac inflammatory activity, therefore ruling out the initiation of immunosuppressive therapy. Given the presence of cardiomyopathy without lupus inflammatory activity in PET/CT, a coronary artery study was requested, revealing extensive three-vessel coronary disease.

Discussion and Learning Points: SLE is a systemic autoimmune disease that can affect any organ in the human body. Cardiac involvement is frequent, and depends on multiple variables, including the antibody profile. A triple positivity for antiphospholipid antibodies is often associated with cardiovascular involvement, as shown in our case. It can present itself as myocarditis, but more frequently as coronary heart syndrome, therefore it is important to include non-inflammatory syndromes within the broad differential diagnosis and to not underestimate the cardiovascular risk of these patients.

Keywords: *systemic lupus erythematosus, cardiovascular risk, atherosclerosis*

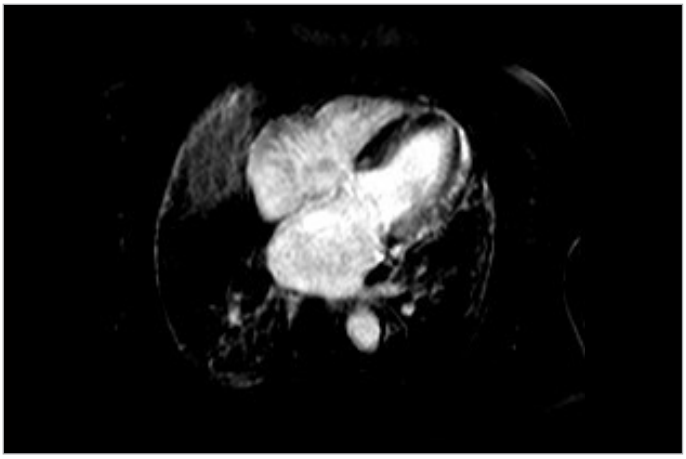


Figure 1. *Cardiovascular magnetic resonance imaging. Four chamber view.*

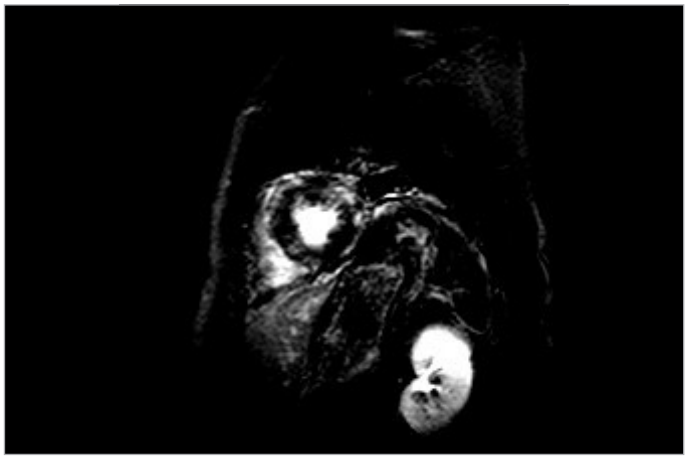


Figure 2. *Cardiovascular magnetic resonance imaging. Short axis view.*

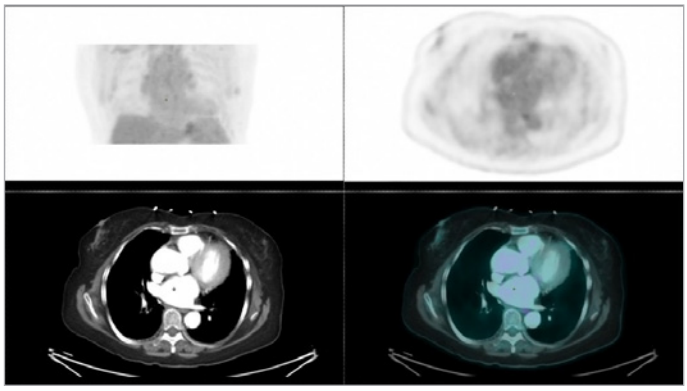


Figure 3. *PET/CT without cardiac inflammatory activity.*

[Abstract:1946]

RESCUE TREATMENT WITH INTRAVENOUS ANAKINRA INFUSION IN STILL'S DISEASE: A CASE REPORT

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[5] Department of Anesthesiology and Reanimation, Baskent University Faculty of Medicine, Ankara, Turkey

Case Description and Diagnostic Pathways: We present a 32-year-old man successfully treated with anakinra after developing macrophage activation syndrome (MAS) associated with adult-onset Still's disease (AOSD). Previously diagnosed with AOSD, patient discontinued his medications and was admitted to the hospital with complaints of sore throat, fever, drowsiness, double vision and severe headache that lasted for about 3 days. Cranial MRI revealed hyperintense lesions in the pons, basal

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ganglia and left parietal region. (Figure 1). Laboratory tests showed, CRP 263 mg/L, ferritin 1204 ng/mL, fibrinogen 601 mg/dL. No infection focus was found in microbiological examinations and cultures and thoraco-abdominal CT showed no signs of infection. Empirical treatment with ceftriaxone and acyclovir was initiated. A preliminary diagnosis of MAS-related encephalitis led to treatment of pulse corticosteroid (1 g methylprednisolone for 10 days), 1 g daily IVIG for 5 days. On the third day of this treatment, the patient developed progressive deterioration in consciousness. Consequently, a three-day infusion of anakinra at 0.5 mg/kg/hour (total 700 mg/d) was started. Significant improvement in the patient's condition was observed at the 12th hour after anakinra infusion. A follow-up brain MRI two months post-treatment revealed complete disappearance of lesions and edema (Figure 2). The patient is currently under monitoring with subcutaneous anakinra (2x100 mg), cyclosporine (2x25 mg), and methylprednisolone.

Discussion and Learning Points: AOSD associated MAS is a rare but life-threatening complication. In this case, AOSD-associated MAS resolved with steroid and anakinra treatment emphasizing. The critical role of IL-1 in the pathogenesis of Still's disease.

Keywords: *adult-onset Still's disease, aacrophage activation syndrome, anakinra*

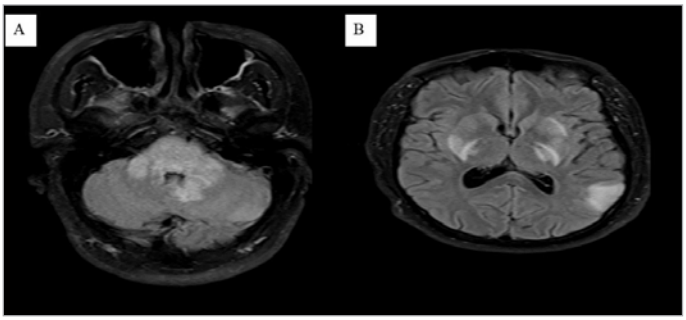


Figure 1. *1A: In the axial FLAIR-weighted sequence, hyperintense lesions were observed in the pons, with expansile features extending to the cerebellar peduncle on both sides and the cerebellar hemisphere on the left. 1B: Hyperintense lesions were observed in the cortex-subcortical white matter of the bilateral basal ganglia capsule interna and left parietal region.*

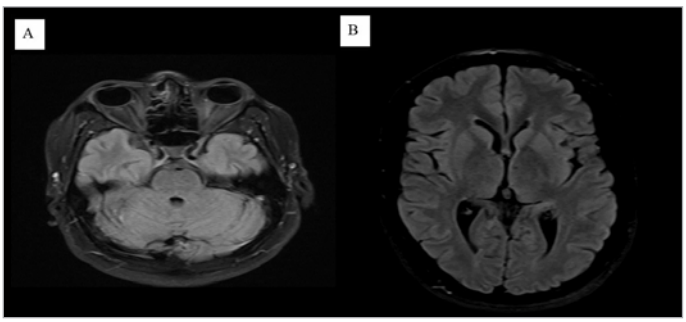


Figure 2. *2A: It was observed that the lesion observed at the pons level disappeared in the control brain MRI 2 months after the treatment. 2B: It was observed that the lesions observed in the basal ganglia, left capsule interna and left parietal region disappeared.*

[Abstract:1949]

"CHLOROQUINE MACULOPATHY": IS THIS PHENOMENON GIVEN ENOUGH ATTENTION?

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Chloroquine maculopathy is a not so rare complication of long-term use of chloroquine preparations in the treatment of rheumatological diseases. Its early detection is of exceptional importance for timely discontinuation of the drug in order to prevent irreversible vision damage.

The aim of this article was to present a case of a 62-year-old patient with bull's eye maculopathy as a result of long-term use of chloroquine due to systemic lupus erythematosus.

The patient presented with blurring and distortion of the pericentral vision. She was ophthalmologically evaluated with fundoscopic examination, autofluorescence, perimetry, optical coherent tomography which together with the medical history established a diagnosis of chloroquine maculopathy. Chloroquine therapy was discontinued and the patient is regularly followed by a Rheumatologist and an Ophthalmologist, with no disease progression so far.

Keywords: *chloroquine, maculopathy, Bull's eye*

[Abstract:1951]

A CASE REPORT ON POLYMYOSITIS IN A PATIENT WITH ESTABLISHED SYSTEMIC LUPUS ERYTHEMATOSUS

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[2] PHI General Hospital Prilep, Prilep, North Macedonia

Introduction: The predisposition to develop an autoimmune disease is thought to be controlled by genetic factors, while the environmental factors are of secondary importance. A female patient who was physically overwhelmed at work, marked by the production of different subsets of autoantibodies during a short period of exacerbation is reported. Her initial presentation as systemic lupus erythematosus (SLE) shifted to the onset of polymyositis (PM) with production of elevated liver enzymes and high creatine kinase levels that tightened the patient in a bed-dependent position.

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The latter titer seemed to change during this relapse and disappeared when myositis resolved.

Aim: This case with autoantibodies shifting followed by the variable clinical manifestations, from muscle weakness, extreme hair loss, polyarthralgia, lower gastrointestinal bleeding and inability to move– argues that environmental factors play an important role in autoimmunity.

Materials and Methods: Different laboratory tests and imaging techniques were conducted since the first patient's hospitalization at the gastroenterohepatology department. The results that showed severe liver damage and lower gastrointestinal bleeding at the beginning were misleading and only lost the time needed to diagnose and treat the real disease that started to show.

Keywords: *polymyositits, SLE, overlap*

[Abstract:1955]

MORE THAN JUST HIVES

Ana María Aldea Gamarra, Fernando Jiménez Segovia, Cristina Lavilla Olleros, Cristina Ausín García, María Victoria Villalba García, Luis Antonio Álvarez Sala Walther, María Barrientos Guerrero

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Case Description: We present a 57-year-old woman with a history of asthma, rhinoconjunctivitis and sinus polyposis in allergy follow-up for suspicion of chronic urticaria in treatment with omalizumab and prednisone.

The patient arrived at the Emergency Room due to persisting symptoms, requiring prednisone almost daily. Physical examination showed facial edema and lumped lesions in the abdomen and neck. She also reported recurring impactions of liquids and solids, arthralgia that improved with prednisone and intense asthenia, along with night sweats.

Blood tests revealed severe eosinophilia of 20,000 cells/uL, positive anti-nuclear antibodies 1/320, negative antineutrophil cytoplasmic antibodies and elevation of lactate dehydrogenase and vitamin B12.

Endoscopic study showed eosinophil infiltration at the level of proximal oesophagus without meeting the criteria for eosinophilic gastroenteritis. The biopsy of the skin lesions did not reveal vasculitis and a bone marrow analysis was performed, being negative for chronic eosinophilic leukemia.

Clinical Hypothesis: Hypereosinophilic syndrome (HES) was considered.

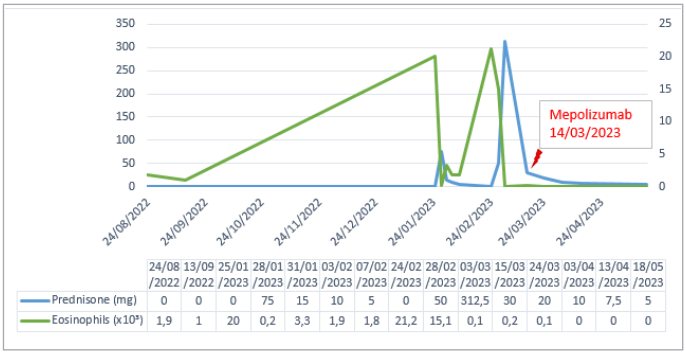
Diagnostic Pathways: A thorough anamnesis, autoimmunity tests and several biopsies discarded other causes of eosinophilia such as reactive, parasitic infections, the drug rush with eosinophilia and systemic symptoms syndrome (DRESS), eosinophilic gastrointestinal diseases, eosinophilic granulomatosis with polyangiitis (EGPA) as well as myeloid/lymphoid neoplasms.

Discussion and Learning Points: HES consists of a group of disorders characterized by accumulation of eosinophils in peripheral blood (>1.5 x 10^9/l) accompanied by organ damage.

We must exclude secondary causes to reach the right diagnosis.

The fundamental treatment is corticosteroids, although other biological agents such as mepolizumab has proved to be an effective treatment.

Keywords: *hives, hypereosinophilic syndrome, mepolizumab*



	24/08 /2022	13/09 /2022	25/01 /2023	28/01 /2023	31/01 /2023	03/02 /2023	07/02 /2023	24/02 /2023	28/02 /2023	03/03 /2023	15/03 /2023	24/03 /2023	03/04 /2023	13/04 /2023	18/05 /2023
Prednisone (mg)	0	0	0	75	15	10	5	0	50	312,5	30	20	10	7,5	5
Eosinophils (x10^3)	1,9	1	20	0,2	3,3	1,9	1,8	21,2	15,1	0,1	0,2	0,1	0	0	0

Figure 1. *Evolution between dose of prednisone and eosinophilia.*

TIME	EOSINOPHILS (x10^3)	PREDNISONE (mg)
24/08/2022	1.9	0
13/09/2022	1	0
25/01/2023	20	0
28/01/2023	0.2	75
31/01/2023	3.3	15
03/02/2023	1.9	10
07/02/2023	1.8	5
24/02/2023	21.2	0
28/02/2023	15.1	50
03/03/2023	0.1	312.5
15/03/2023	0.2	30
24/03/2023	0.1	20
03/04/2023	0	10
13/04/2023	0	7.5
18/05/2023	0	5

Table 1. *Evolution between dose of prednisone and eosinophilia.*

[Abstract:1957]

HERPETIC RETINITIS IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Servico Medicina 2.3, Hospital Santo António dos Capuchos, Lisbon, Portugal

A 30-year-old woman, diagnosed with systemic lupus erythematosus (SLE) when she was 16 years old, had inflammatory signs on her right eye during her holidays through Europe. Back in Portugal, she initiated topical eye treatment aimed to treat bacterial conjunctivitis in the Emergency Room (ER). On the next day, she returned to the ER, she took valacyclovir for presumed herpes infection. Four days later, with worsening pain and noticeable reduced visual acuity, she was hospitalized to be given

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parental therapy. A physical exam showed edema, redness, and inability to open her right eye. An ophthalmologic exam revealed fixed mydriasis, acute retinal necrosis, and conjunctival hyperemia. In the aqueous humour herpes simplex virus was isolated. Head CT showed pre-septal cellulitis and optic nerve neuritis affecting the oculomotor, facial, and vestibular nerves. Despite several weeks of high doses of intravenous antiviral and antibacterial medication, she was discharged with a loss of visual acuity.

Recently, she had been prescribed anifrolumab for SLE, a type I interferon receptor blocker. However, this was discontinued for its potential infection risk.

Acute retinal necrosis is associated with varicella-zoster, herpes, cytomegalovirus, and Epstein-Barr virus infection, particularly in individuals taking immunosuppressant drugs. Additionally, clinical trials of anifrolumab showed a higher infection risk by herpes zoster virus (12.2%) compared to placebo (7.2%).

Keywords: *acute retinal necrosis, systemic lupus erythematosus, immunosuppressive therapy*

[Abstract:1975]

SARCOIDOSIS IN A PORTUGUESE COHORT

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Sarcoidosis is a multisystemic granulomatous disease with variable clinical manifestations, organ involvement and course. The aim of this study was to assess epidemiologic and clinical features, work-up and treatment in patients with sarcoidosis from a tertiary hospital in Portugal.

A retrospective cohort analysis of patients with sarcoidosis followed from 2010-2022 at an Internal Medicine department was performed, following an individual medical record review.

We identified 56 patients with sarcoidosis (mean age of 54.2±15.9 years; 60.7% females and 80.4% caucasian), in which 23.2% were active/previous smokers, 10.7% had an history of occupational exposure and 16.1% had auto-immune comorbidities. Most patients had an increased angiotensin-converting enzyme and sedimentation rate. Thoracic involvement occurred in 85.7% of patients, and among extrathoracic manifestations, the most frequent was ophthalmological disease (35.7%), followed by cutaneous involvement (30.4%), musculoskeletal (23.3%), renal and gastrointestinal (3% each). Non-specific symptoms such as fatigue (37.5%), weight loss (14.3%) and fever (12.5%) were frequent. Regarding disease presentation, uveitis was the most common form (32.1%), followed by erythema nodosum (16.1%) and constitutional symptoms (8.9%). Only 8.9% of patients had no need for systemic treatment – systemic corticosteroids were used in 91.1% of cases, other immunosuppressors in 62.5% (tumor necrosis factor inhibitors in 17.9% of cases). The mortality rate was of 8.9%, though none of the deaths were related to disease activity. Sarcoidosis remains a diagnostic challenge due to the varied clinical manifestations and absence of pathognomonic disease markers, and cohort studies are essential to raise awareness to this diversity and improve diagnostic acuity.

Keywords: *sarcoidosis, extrathoracic involvement, clinical presentation*

[Abstract:2005]

INTERSTITIAL LUNG DISEASE AS THE FIRST MANIFESTATION OF SYSTEMIC SCLEROSIS: FROM DIAGNOSIS TO TREATMENT

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A 70-year-old man, ex-smoker with a history of diabetes and psoriasis, was admitted to the hospital for dyspnea and productive cough for two months. He was febrile, with tachypnea, scattered lung crackles, and no peripheral edema. Laboratory tests presented hypoxemic respiratory failure, C-reactive-protein of 240 mg/L. Chest computed-tomography revealed subpleural reticulation, honeycombing areas, and traction bronchiectasis concerning pulmonary fibrosis, besides ground-glass infiltrates, reflecting a possible infectious exacerbation [Fig1]. Common respiratory viruses, urinary pneumococcal and legionella antigens, and blood cultures were negative. Bronchofibroscopy revealed intense neutrophilic and mild eosinophilic alveolitis, no isolation of microbiological agents, and was negative for malignancy. Corticosteroid therapy (CCT) 0.75 mg prednisolone/kg and empirical antibiotic therapy were started, with clinical and analytical improvement. However, after discharge he presented a new clinical and imaging worsening. The immune study revealed positivity for antinuclear antibodies (ANA) 1:1280, anti-centromere (ACA) A and B, and RNA-polymerase-3 (Pol3), culminating in the diagnosis of pulmonary involvement by systemic sclerosis (SSc). He restarted CCT 0.5 mg prednisolone/kg. After the authorization, he initiated treatment with mycophenolate mofetil, increased up to 3 g/day, and nintedanib, with clinical stability, allowing progressive reduction of CCT.

Interstitial lung disease (ILD) encompasses a wide group of diseases that cause pulmonary fibrosis. It is a diagnostic challenge and it is often progressive and irreversible. ILD is one of the

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manifestations of SSc, but it is rare to be the first manifestation. It is more associated with positivity for anti-topoisomerase-I and anti-RNP-U3 antibodies, not for ACA and Pol3 as in this case. Immunosuppressive treatment should be started as early as possible.

Keywords: *systemic sclerosis, interstitial lung disease, mycophenolate mofetil*

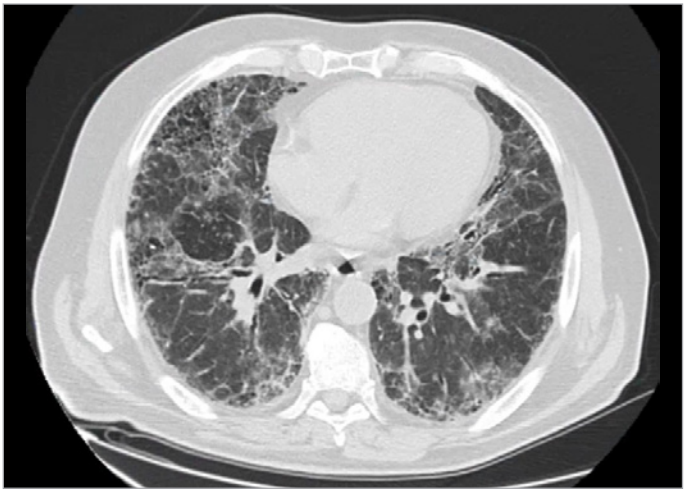


Figure 1. *Subpleural reticulation, honeycombing areas, and traction bronchiectasis concerning pulmonary fibrosis, and ground-glass infiltrates, reflecting a possible infectious exacerbation, on chest-computed tomography.*

[Abstract:2011]

RECURRENT PANUVEITIS; WHO IS THE CULPRIT? A CASE REPORT

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Uveitis refers to inflammation of the eye. Tuberculosis origin is rare. The occurrence of uveitis during an inflammatory rheumatism raises the possibility of infectious uveitis due to immunosuppression. This case report presents a 31-year-old patient known for JIA since childhood, labelled as rheumatoid arthritis in adulthood and irregularly treated, with a history of cutaneous and ongoing lymph node tuberculosis, was referred for the management of recurrent bilateral panuveitis since the age of 16. Ophthalmologic examination revealed a red and painful right eye with decreased visual acuity. Angiography and OCT showed signs of active panuveitis in both eyes, with choroid-retinal lesions suggestive of retinal tuberculosis. Thoracic CT did not show miliary patterns. Joint examination revealed typical deformities of rheumatoid arthritis with a high DAS28 activity score. The diagnosis of tuberculous and rheumatoid panuveitis was suspected based on the patient's history, concurrent active lymph node tuberculosis, and findings from angiography and OCT. Treatment involved a regimen of anti-tuberculosis drugs and corticosteroids with a gradual tapering schedule and alternative options for anti-TNF therapy.

This case underscores an unusual connection between tuberculosis, rheumatoid arthritis, and panuveitis, urging clinicians to consider tuberculous panuveitis even in rheumatoid arthritis cases, supported by comprehensive medical history, clinical, laboratory, and radiological evidence.

References:

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2. S. Lechtman, V. Queyrel, C. Maschi, J.G. Fuzibet, P. Gastaud, S. Baillif-Gostoli, N. Tieulie Critères diagnostics des uvéites tuberculeuses https://doi.org/10.1016/j.revmed.2014.10.150

Keywords: *panuveitis, tuberculosis, Juvenile Idiopathic Arthritis, rhumatoid arthritis*

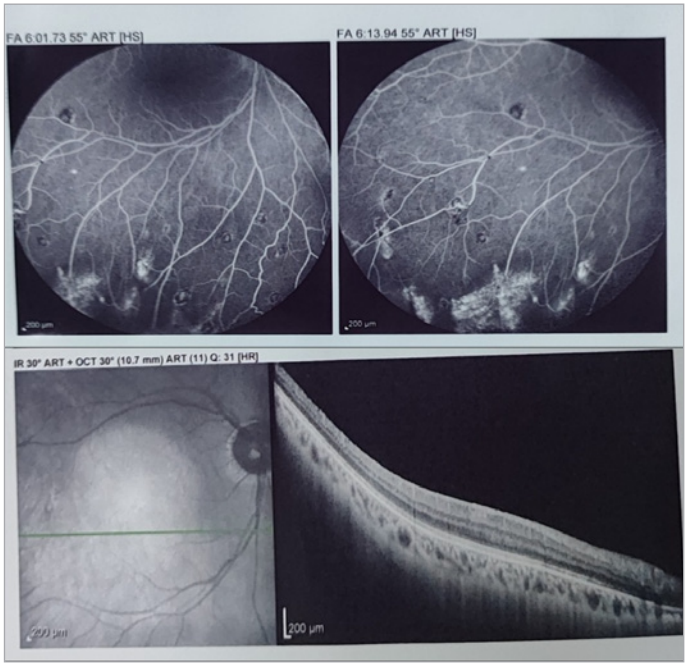


Figure 1. *Angiography and OCT result.*
Panuveitis, with choroid-retinal lesions suggestive of retinal tuberculosis.

[Abstract:2013]

SARCOPENIA IN WOMEN WITH RHEUMATOID ARTHRITIS

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Background: Rheumatoid arthritis is a common rheumatic disease with high prevalence of sarcopenia, formed by low functional

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activity, chronic inflammation, nutritional disorders. A number of medicaments are used. EWGSOP2 criteria are not fully suitable for patients with RA because they often have pain and joint deformations and have difficulties in functional tests performing. That is why instrumental methods may be useful in identifying sarcopenia. The aim of our study was to evaluate body composition parameters and its association with SARC-F questionnaire results and functional test in women with rheumatoid arthritis.

Materials and Methods: Sixty-five women with rheumatoid arthritis (2010 ACR/EULAR criteria) were included. Body composition was carried out by dual-energy absorptiometry. SARC-F questionnaire and 5 times getting up from a chair test were carried out for screening of sarcopenia presence.

Results: SARC-F result more than 3 was revealed in 34 (52.3%) patients. The time to rise from a chair was longer in these patients ($p<0.001$). 12 (18.5%) patients were unable to complete the test due to joint pain. Sarcopenia, evaluated by low appendicular muscle index (AMI≤5.5 kg/m^2), was found in 12 (18.5%) patients and did not correlate with SARC-F results. Four patients from 34 had low AMI and positive SARC-F result, but 8 patients from 31 had low AMI and normal SARC-F result.

Conclusions: Sarcopenia assessment using questionnaires and functional tests may have limitations in RA patients and didn't correlate with DXA results. That is why DXA should be used in these patients to detect decreased muscle mass.

Keywords: rheumatoid arthritis, sarcopenia, SARC-F, DXA

[Abstract:2023]

FROM A COLLATERAL VENOUS CIRCULATION TO AN ANGIO-OCULO- BEHÇET: ABOUT A CLINICAL CASE

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Introduction: The term "vasculo-Behçet" used to define the forms of Behçet's disease where vascular impairment dominates the picture. Arterial impairment is less common than venous impairment. The oculo- Behçet touch the different segments of the eye whose posterior uveitis is of poor prognosis.

Materials and Methods: We report the case of a 41-year-old non-smoking man.

Results: The disease history, 2 years earlier, was marked by the gradual installation of a weakness in the two lower limbs with intermittent claudication taken as a discal hernia with sciatalgia, and subsequent appearance of a collateral venous circulation in the abdominal wall.

Clinical Examination: Recurrent oral aphthoses, scars of genital aphthoses, large collateral venous circulation in the abdomen, coldness of the lower limbs with a pulse not felt, blurry vision with a painful red eye. TA was at 180\110 mmhg. HLA B51 was positive. Abdominal angio-scanner revealed thrombotic disease responsible of extensive total obstruction of the abdominal aorta, left common iliac artery and subtotal left renal artery ostium with signs of compensation by supplement circulation.

Findings: OS sat a posterior uveitis with hyalitis, macular edema, occlusive vasculitis and arterio-venous shunts. The diagnosis of Behçet's disease is taken according to the new 2013 criteria. Patient was put under bolus corticoids, then withdrawed gradually with azathioprine. Pre-therapeutic assessment of a biotherapy found a quantiferon positive, so the patient was put on adalimumab at 21 day of anti-TBC treatment.

Conclusions: Angio-oculo-Behçet impairments condition the prognosis of Behçet's disease.

Keywords: Behçet's disease, ocular impairment, vascular impairment

[Abstract:2026]

PROBABLE CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME - A RARE CASE REPORT

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[2] Department of Cardiology, Maltepe University, Istanbul, Turkey

Case Report: We present a 55-year-old male patient with known rheumatoid arthritis, first presenting to the emergency department with chest pain. He was diagnosed with hyperacute, ST-elevated myocardial infarction.

Trans-thoracic echocardiography revealed left ventricle and atrium dilatation, left ventricle systolic disfunction and 1.9x1.9 cm thrombus at apex. Coronary angiography showed acute coronary syndrome with thrombus starting from distal left main coronary artery, through left anterior descending artery (LAD), TIMI 2-3 flow, thrombus at mid circumflex artery.

Tirofiban therapy was started. There was no evidence of thrombus dissolution on control angiography, so the patient was started on tPA (tissue type plasminogen activator) infusion. On day 1 of tPA infusion, the patient developed confusion; left parietal hypodense lesion, hypodensity of right anterior and posterior Watershed areas was seen on cranial CT, which were suggestive of acute ischemic cerebrovascular incidence. On day 7, therapy was changed to oral warfarin, acetylsalicylic acid 100 mg and enoxaparin sodium 6000 IU. Purpuric lesions on bilateral legs were remarkable during physical examination 4 weeks after the first admission.

Clinical Hypothesis: Probable catastrophic antiphospholipid syndrome

Diagnostic Pathways: Antiphospholipid antibodies resulted negative, whereas lupus anticoagulant was detected as strongly positive twice, consistently with the diagnosis of probable catastrophic antiphospholipid syndrome (CAPS).

Discussion: CAPS is a life threatening and infrequent form of antiphospholipidsyndrome.Itpresentswithmultiplemicrovascular or large vessel thrombosis developing simultaneously or in a short period of time in multiple vascular beds and result in multiple

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organ failure. Early and prompt identification is necessary to prevent worst outcomes.

Keywords: *catastrophic antipospholipid syndrome, thrombosis, Lupus anticoagulant*



Figure 1. *LMCA Thrombus on day 0.*

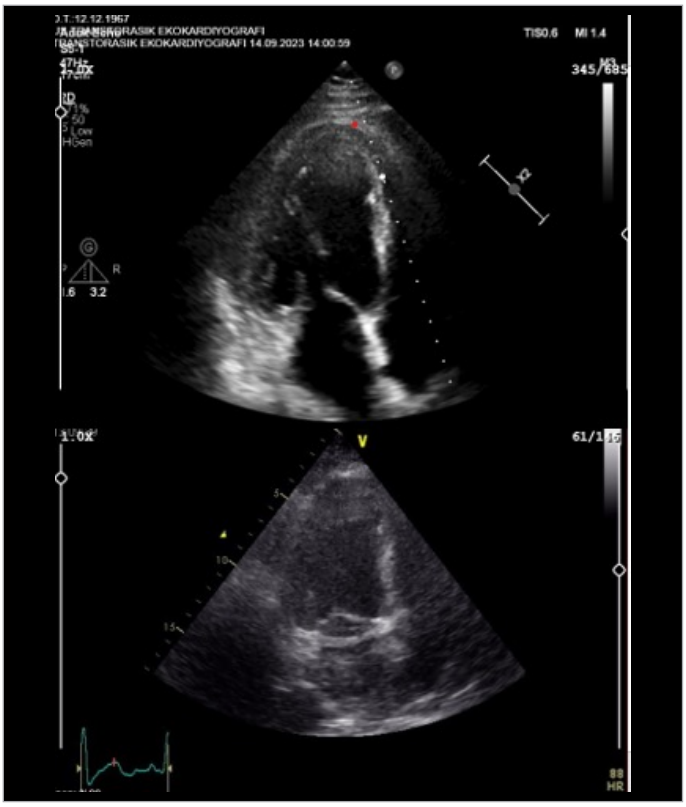


Figure 2. *Apical thrombus seen on echocardiography 1 week and 2 months after first admission.*

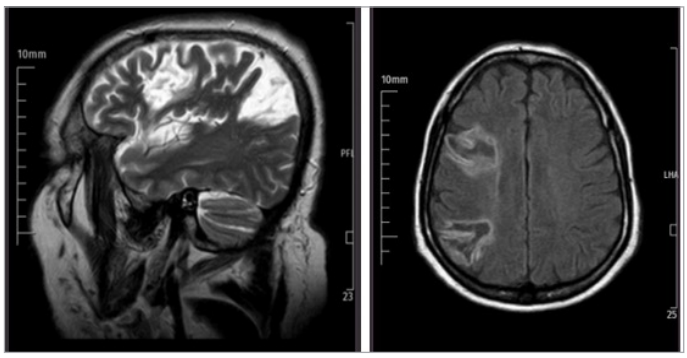


Figure 3. *MRI- Acute ischemic infarctus of watershed areas.*



Figure 4. *LMCA thrombus is seen dissolved at control angiography before discharge.*

[Abstract:2027]

ANTERIOR UVEITIS WITH LARGE VESSEL VASCULITIS

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Case Description: A 59-year-old woman with hypertension, diabetes mellitus and previous history of ischemic stroke (CVA) was admitted with difficulty in speaking and walking. One month earlier she was started on methotrexate(MTX) for uveitis with pre-diagnosis of Behçet's disease (BD). She declared the usage failure in MTX as daily and was hospitalized with MTX toxicity.

Clinical Hypothesis: Large vessel vasculitis (LVV) should be considered in ocular involvement at older age.

Diagnostic Pathways: Physical examination showed no mucositis or joint abnormality. Laboratory showed pancytopenia caused by MTX toxicity and elevated acute phase reactants. Calcium folinate was started with oral folate combination. She denied

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recurrent oral aphte, genital ulcers, folliculitis, arthritis, erythema nodosum. On ocular examination posterior synechia was found addressing previous uveitis attack. Owing her older age, ocular involvement was not associated with BD and vascular screening was requested for LVV and showed wall thickening of the aorta and its branches, suggesting active inflammation, and total occlusion of left common carotid artery. With diagnosis of LVV, 40 mg/day methylprednisolone was given and she well responded.

Discussion and Learning Points: LVV is the inflammation of the aorta and its branches and most common primary vasculitis in elderly patients.

Although leading etiology of posterior uveitis is BD, it is usually affects younger and male patients in first 5 years of BD. In older patients with unexplained acute phase elevation or vascular involvement as CVA differential diagnosis should include LVV. To proper evaluation vascular imaging techniques are useful. Early and effective immunosuppression with corticosteroids is mainstay of the treatment.

Keywords: *Behçet's disease, vasculitis, large vessel vasculitis, uveitis*

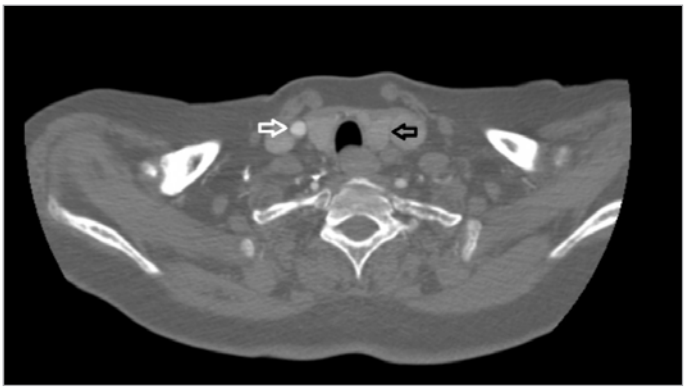


Figure 1. *On computerized tomography angiography (CTA) of patient, total occlusion of left common carotid artery (black arrow) revealing vasculitis with vessel wall thickening and normal vascular appearance and contrast filling of right common carotid artery (white arrow) were observed.*

[Abstract:2041]

IDIOPATHIC ACQUIRED ERYTHROBLASTOPENIA MIMICKING HEPATO-SPLENIC LYMPHOMA

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Background: Idiopathic acquired erythroblastopenia (IAE) is characterized by elective absence of erythroblasts in the marrow. They are associated generally with viral infections, malignant hemopathies, or auto-immune diseases.

We report a case of IAE mimicking hepato-splenic lymphoma.

Case Report: A 19-year-old female patient was admitted for severe anemia in 3.9 g/dl of hemoglobin. There was neither icterus nor bleeding.

Physical examination reported paleness, functional systolic heart murmur, painless, enlarged liver, and homogeneous splenomegaly with a coastal overhang of 3 cm. There were no enlarged lymph nodes or cutaneous involvement. The hematologic evaluation showed normocytic anemia in 3.9 g/dl. The rate of reticulocytes was 14700/mm^3. There were no biological signs of hemolysis. Chest radiography did not reveal mediastinal extension. A bone marrow smear showed erythroblastopenia with a 14% of lymphocytosis.

The histopathological examination of the bone and liver draining biopsies showed interstitial and nodular lymphoid marrow infiltration occupying 60% of the marrow surface constituted by small-sized lymphocytes and a discreet hepatic infiltration by lymphocytes. In front of the absence of monoclonal character in the immune-histochemical study, the diagnosis of hepatosplenic lymphoma was not retained. The search for hemoglobinopathy, viral causes, and autoimmune diseases was negative.

The diagnosis of IAE was then retained and the patient was treated with corticosteroids. The clinical and biological outcome was favorable with a spectacular improvement of the hemoglobin which passed to 11 g/dl.

Conclusions: Idiopathic acquired erythroblastopenia, a real autoimmune disease of the erythroblast, can be confused with many other diseases, particularly malignant diseases.

Keywords: *erythroblastopenia, hematology, lymphoma, auto-immune disease*

[Abstract:2063]

ROLE OF SUBTLE SYMPTOMS AND WHOLE-BODY PET-CT SCAN IN THE DIAGNOSTIC WORK-UP OF A WOMAN WITH AORTIC ANEURYSMS

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A 73-year-old woman was admitted to our unit with a four-week history of dyspnea, myalgias and polyarthralgias. Her past medical history was significant for bio-prosthetic aortic valve replacement, thoraco-abdominal aneurysm treated with endovascular repair (four years prior to the admission) arterial hypertension, permanent atrial fibrillation, chronic disseminated intravascular coagulopathy and osteoporosis. Presenting complaints included new onset headache of the left region of the face with homolateral flushing, carotidynia and tenderness of the temporal area. On clinical examination, she appeared dyspnoic, with limbs' movement limitation due to proximal muscle pain, and polyarthralgias of the distal joints. Laboratory findings showed neutrophilia, C-reactive protein of 22 mg/L, normal erythrocyte

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sedimentation rate, hyperfibrinolysis and hypofibrinogenemia.
As the suspicion of polymyalgia rheumatica (PR) and giant cell arteritis (GCA) was raised, we performed laboratory test for autoimmune vasculitis (p-ANCA, c-ANCA) and for non-vascular inflammatory disease (ANA, ENA, FR), that resulted negative. Interleukin-6 serum levels were 8.3 pg/mL. Two sets of blood cultures resulted negative. Doppler ultrasound of temporal artery and main arteries didn't show inflammatory changes. Whole-body PET-CT scan showed increased tracer uptake in the thoracic aorta (maximum SUV 4.87). A presumptive diagnosis of long-standing, unrecognized PR with GCA was done. Once we had excluded differential diagnosis, GCA was plausible according to the latest classification criteria. In details, the patient satisfied three clinical criteria, one laboratory criterion and one imaging criterion, scoring 11 points. Considering the history of osteoporosis limiting high dose steroid administration, the patient is currently under evaluation for treatment with tocilizumab.

Keywords: *rheumatological disease, giant cell arteritis, polymyalgia rheumatica*

[Abstract:2064]

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (GEPA) AND LOEFFLER'S ENDOCARDITIS. APROPOS OF A CASE

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We report the case of a 44-year-old woman, smoking 10 cigarettes/day. Her personal history included dyslipidemia, overweight, Graves' disease, bronchial asthma treated with bronchodilators, sinusitis and rhinitis. She came to the emergency for having progressive dyspnea for 3 weeks under minimal effort, increased edema in the lower limbs, orthopnea and paroxysmal nocturnal dyspnea. In the latest week she reported hypoesthesia in left lower limb. No motor deficit. No arthralgias or arthritis or other symptoms of interest. She was afebrile with no infectious symptoms. Physical examination highlighted the presence of edema with pitting in lower limbs. Electrocardiogram ST in II, III, aVF, V5-V6, showed negative T V5-V6. Analytically: eosinophilia, thrombopenia, D-dimer 1785 ng/mL and troponins 1770 ng/L. Positive anti-cytoplasmic antibodies IgG anti-myeloperoxidase p-ANCA. Angio-CT ruled out pulmonary thromboembolism. Given the asthmatic history of the patient and the analytical data obtained, the diagnosis of eosinophilic granulomatosis with polyangiitis was established. The echocardiogram showed a pattern of myocardial restriction, apical hypertrophy, biauricular dilatation and thrombi in the ventricular wall, diagnosis of Loeffler's endocarditis. Treatment was started with three 1-gram boluses of methylprednisolone and cyclophosphamide in 500 mg boluses for three days. After treatment, there was considerable decrease in troponins, eosinophils and NT-proBNP. On MRI, there was biventricular endomyocardial fibrosis, with left ventricular apical thrombus. Follow-up in consultation with adherence to treatment with cardiac remodeling and asymptomatic. The following year, she abandoned treatment and went to the emergency with ischemic lesions established in the superficial and distal territories of the middle cerebral artery, posterior and posteroinferior cerebellar: GEPA recurrence and secondary complication Loeffler's endocarditis and thromboembolic stroke.

Keywords: *GEPA, Loeffler endocarditis, autoimmunity, eosinophils*

[Abstract:2074]

INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: NINE CASES

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Background: Diffuse interstitial lung disease (ILD) with autoimmune features (interstitial pneumonia with auto-immune features or IPAF) is an overlap between idiopathic and associated to connectivites. It is a new entity that is currently characterized.
Patients and Methods: We performed a retrospective descriptive study between 2015 and 2023, collecting 160 cases of DIP. Nine cases of IPAF were indentified (5% of PID). The diagnosis of IPAF was based on the American Thoracic Society and the European Respiratory Society criteria.
Results: Eight of our patients were women. The mean age was 55 years. IPAF was revealed by respiratory symptoms in 7 cases. The main respiratory signs were cough (9 cases). Extra-respiratory manifestations were present in 7 cases. These were cutaneous signs (6 cases), articular (5 cases), xerostomia (4 cases), xerophthalmia (3 cases) and Raynaud's phenomenon (1 case). Periungual capillaroscopy performed in 6 cases revealed non-specific abnormalities in 4 cases. The immunological abnormalities observed were antinuclear antibody positivity antinuclear antibodies (2/3 of cases), rheumatoid factor (4 cases) and anti-CCP (2 cases). The anti-ENA antibodies were positive in 3 patients. Corticosteroid therapy was prescribed in six patients, with immunosuppressive therapy in one patient. The overall evolution was improvement (3 cases), worsening (3 cases) and stabilization (3 cases). The mean follow-up was three years. There was a significant correlation between cutaneous signs during IPD and IPAF (p=0.047).
Conclusions: Further prospective studies are needed to refine IPAF criteria. The current criteria cover a heterogeneous group of patients making impossible to identify a common profile.

Keywords: *auto-immune disease, connective tissue disease, interstitial lung disease*

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[Abstract:2131]

AMYLOIDOSIS AND CASTLEMAN'S DISEASE ASSOCIATED WITH H.SUPURATIVA

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[3] Department of Dermatology Dokuz Eylul University, Izmir, Turkey

Amyloidosis is a fatal and incurable disease of abnormally folded proteins, both intracellular and extracellular, causing loss of function of organs and systems, of which at least 35 different types have been identified so far, and its progression is often linked to the underlying primary condition associated with chronic inflammation that initiates AA amyloidosis.

Hidradenitis suppurativa is relatively a rare cause of amyloidosis, cause large scale of morbidity and discomfort, often incurable and yet its systemic persistent inflammatory status often overlooked.

We present the case of a 59-year-old male who suffered from HS for 20 years and had a stage 3 (KDIGO) chronic renal failure with unknown reason. He had been hospitalized for planning anti-TNF biologic treatment to HS and mild transaminase elevation was detected. When other prominent causes had been ruled out, liver biopsy was performed, which revealed amyloidosis.

Tissue biopsy from kidney could not be obtained due to cortex thinness but now 1 g/dL proteinuria allowed us to refer to systemic secondary amyloidosis. Nevertheless abdominal subcutaneous fat biopsy revealed amyloidosis as well.

Patient had progressed to decompensated cirrhosis of the liver and also required renal replacement therapy with automated peritoneal dialysis.

In the course of follow-ups, noticeable and clinically relevant inguinal lymphadenopathy was excisionally biopsied and revealed Castleman's disease; angio follicular lymph node hyperplasia, resulted from the abnormal proliferation of plasma cells and B lymphocytes in lymphoid tissues, which can occasionally be caused by the systemic inflammation ruled by persistent abnormal cytokine, dominantly IL-6 secretion.

Keywords: *Castleman's disease, amyloidosis, hidradenitis suppurativa*

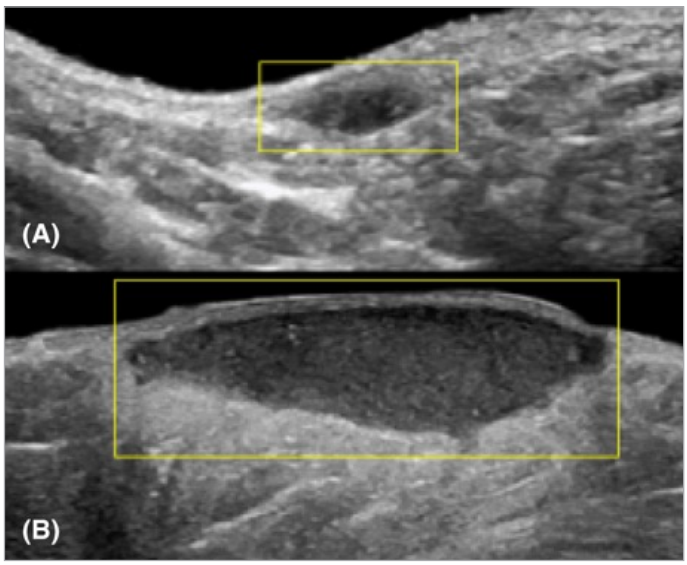


Figure 1. *HS USG images.*
HS lesions nodule (A) and abscess (B).

[Abstract:2136]

THE EFFECT OF INTRAARTICULAR THERAPY WITH HYALURONIC ACID ON THE TIME OF MRI T2 CARTILAGE RELAXATION IN PATIENTS WITH OSTEOARTHRITIS OF THE KNEE JOINTS WITH A SHORT HISTORY OF THE DISEASE

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Purpose: To evaluate the effect of local therapy on the processes of cartilage degradation in knee OA.

Materials and Methods: 7 43–year–old women [35-57] with knee joint OA of stage I-II according to Kellgren-Lawrence and disease duration up to 5 years were included. The average body mass index was 23.75±4.25 kg/m2; the average pain level on a visual analog scale in the knee joint was 32.33 mm [25-40]. Patients received a single intra-articular injection of sodium hyaluronate (GH) 40mg/2ml into the targeted knee joint. MRI T2 mapping of the knee joint was performed before the introduction of GN. Repeated MRI T2 mapping was performed in 2 patients after 6 months, and in 5 patients after 12.

Results: Patients initially did not differ in T2 relaxation time in all layers of cartilage. After 6 months, there was a tendency to decrease the T2 relaxation time: the deep layer was initially 33.55±1.77 ms, after 6 months 31.35±4.45 ms (p=0.58); the surface layer was 38.10±4.24 ms, 34.10±5.09 ms (p=0.48), respectively; total – 34.05±3.04 ms, 31.05±4.59 ms (p=0.52), respectively (Figure 1). In 5 patients, after 12 months, there was a tendency to increase the T2 relaxation time in all layers of cartilage: the deep layer was initially 28.48±1.91 ms, after 12 months 31.08±3.66ms (p=0.2); surface – 34.3±1.29 ms, 36.16±4.23 ms (p=0.37), respectively; total – 30.3±1.49 ms, 31.92±3.38 ms (p=0.36), respectively.

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Conclusions: Effectiveness of Gh drugs is on average 6 months. MRI T2 mapping should be performed 6 months after the use of GN.

Keywords: osteoarthritis, sodium hyaluronate, T2 relaxation time

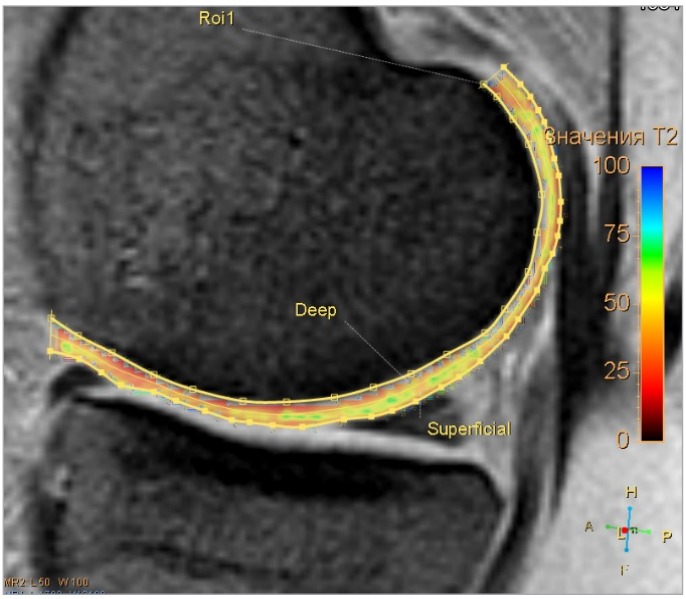


Figure 1. *relaxation time before the introduction of sodium hyaluronate*

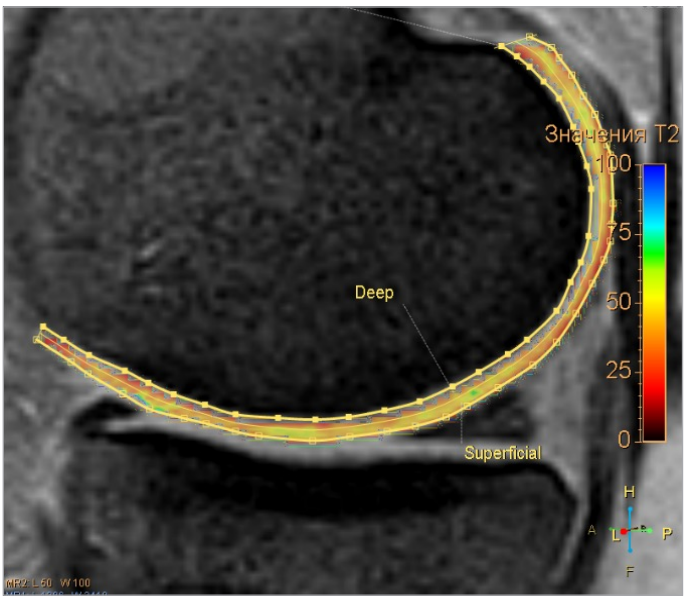


Figure 2. *T2 relaxation time 6 months after administration of sodium hyaluronate.*

[Abstract:2138]

A 30-YEAR-OLD WOMAN WITH DYSPNEA

Víctor Urbaneja Zumaquero, Daniel Ávila Royon, Ángela Eloisa Moliz Molina, Eduardo Manuel Cebreros Cuberos

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Case Description: A 30-year-old woman with no relevant medical history came to the emergency room with pleuritic chest pain accompanied by palpitations and dyspnea of approximately one month's duration. Additionally, she reported alopecia and arthromyalgia in shoulders, elbows, and knees. Upon examination, she exhibited bibasal hypoventilation with mild crackles, arthritis in shoulders, knees, and elbows, and tachypnea, requiring high-flow oxygen therapy.

Clinical Hypothesis: Dyspnea in a patient with possible lupus. The most common causes in this context include infectious etiologies, diffuse alveolar hemorrhage (DAH), and lupus pneumonitis.

Diagnostic Pathways: A complete blood test, including an autoimmune profile, revealed anemia, leukopenia, lymphopenia, and elevated acute-phase reactants. A comprehensive imaging study began with a chest X-ray showing bilateral alveolar infiltrates, followed by an angio-CT revealing multiple ground-glass opacities. Empirical antibiotic therapy was initiated after culture collection. Bronchoscopy ruled out DAH, and with negative cultures, treatment with methylprednisolone pulses and cyclophosphamide was started, leading to progressive improvement in the patient's clinical and radiological condition.

Discussion and Learning Points: Lupus pneumonitis is a rare manifestation of lupus with nonspecific clinical presentation. Given its infrequency, an infectious etiology is initially considered, warranting empirical antimicrobial therapy until ruling out an underlying infection. Treatment involves high-dose methylprednisolone (1 gram/day for 3 days) in addition to immunosuppressive therapy.

Keywords: autoimmune diseases, lupus, lupus pneumonitis, dyspnea

[Abstract:2145]

OBSTRUCTIVE HYDROCEPHALUS IN SARCOIDOSIS: A CASE REPORT

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Introduction: Central nervous system involvement in Sarcoidosis is rare. Hydrocephalus occurs in 6-7% of cases with a poor prognosis. We herein report a case in this regard.

Case Report: A 55-year-old woman was previously diagnosed with systemic sarcoidosis and treated by methotrexate for articular and cutaneous involvement. She presented with a 2-months history of headaches, dizziness, vomiting, amnesia on recent events, blurry vision, urinary incontinence and gait disorders.

On admission, the patient was apyretic. Neurological examination revealed unsteady tandem gait and bilateral diplopia. Other neurologic exams and testing of sphincter tone testing were normal. Her complete blood count; renal, hepatic, phospho-calcic blood tests, C-reactive protein and erythrocyte sedimentation rate were normal.

Magnetic resonance imaging revealed remarkable dilatation of the lateral and third ventricles, periventricular hyper signals,

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enlargement of the Sylvius aqueduct with a slight thickening of its wall and an obstruction at the level of the Magendie foramen with a granuloma of attenuated signal and slight contrast.
Ventriculoperitoneal shunting was conducted. Cerebro-spinal fluid had increased opening pressure of 35 cm H_20. Cytological, bacteriological and biochemical examinations were negative.
Combined treatment with prednisone and cyclophosphamide was started. All neurological symptoms showed marked improvement. Control MRI after six months revealed regression of the periventricular hyper signals and decrease of the measurements of the Magendie foramen obstructive granuloma and the Sylvius aqueduct wall thickening. Ventricular spaces were normal in shape and size.
Conclusions: Hydrocephalus is a rare manifestation in sarcoidosis. Yet, clinicians must suspect it when patients present with common intracranial hypertension symptoms.

Keywords: *sarcoidosis, hydrocephalus, systemic diseases, rare diseases*

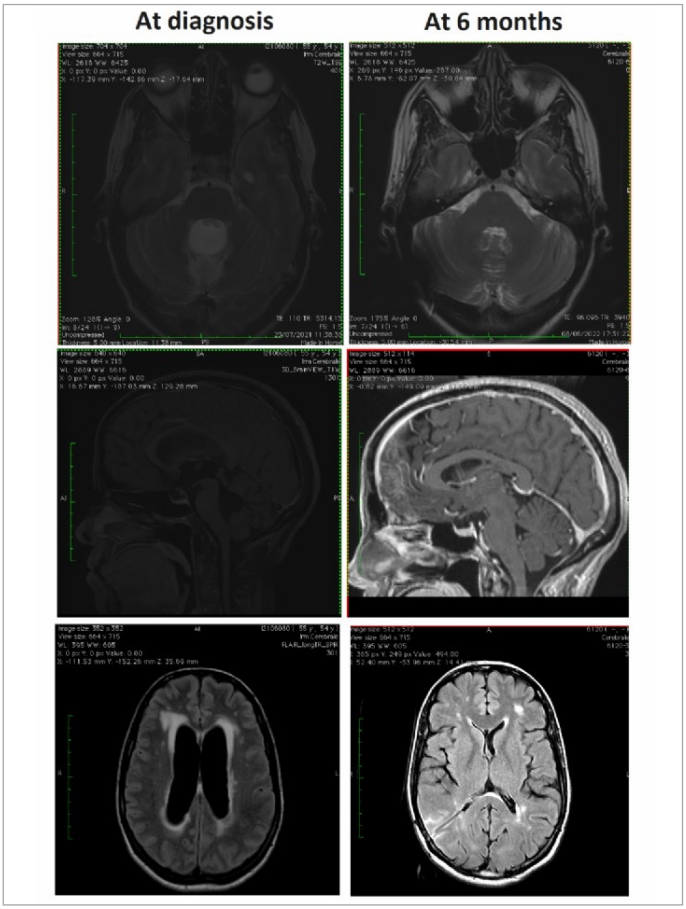


Figure 1. *MRI at diagnosis and after treatment.*

[Abstract:2164]

CEREBRAL INFLAMMATORY PSEUDOTUMOR REVEALING BEHÇET'S DISEASE

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Background: Inflammatory pseudotumor is an uncommon tumor that can involve various organs. Characteristically, the inflammatory pseudotumor is an inflammatory mass leading to manifestations related to its localization. Relatively ubiquitous, this tumor is seldom described in the central nervous system.
We report a case of inflammatory pseudotumor in the central nervous system revealing Behçet's disease.
Case Report: We report the case of a 38-year-old patient, who had bilateral deafness since childhood with difficulty in language and left regressive and recidivated hemiplegia. CT scan showed a right expansive capsule-thalamic process that strongly took the contrast with perilesional edema. The biopsy did not show signs of malignancy. The process disappears with corticosteroids. The diagnosis of Behçet's disease was retained in front of oral and genital aphthosis, skin lesions such as folliculitis, a positive pathergy test, and a massive thrombosis of superior vena cava extended to the stick of the azygos vena with right and left internal jugular thrombosis of 2 veins. Magnetic resonance angiography has eliminated cerebral thrombophlebitis. The ophthalmic exam was normal. The patient was put under anticoagulants and corticosteroids, colchicine, and immunosuppressive treatment (cyclophosphamide followed by azathioprine). The evolution was marked by the obstinacy of the superior vena cava syndrome with the notion of stopping the treatment for 2 months.
Conclusions: The discovery of regressive and recidivated hemiplegia in a young patient must evoke the diagnosis of inflammatory pseudotumor in the central nervous system and Behçet's disease, even in the absence of predisposing ethnic or geographic factors.

Keywords: *pseudotumor, Behçet's disease, central nervous system*

[Abstract:2171]

IMMUNE THROMBOCYTOPENIA AS THE INITIAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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Immune thrombocytopenia is a common hematologic manifestation of systemic lupus erythematosus (SLE) with a prevalence range of 10–25%. However, severe forms of thrombocytopenia occur in only 5% of these patients requiring

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specific management. A 62-year-old man with history of 3 episodes of unprovoked deep venous thrombosis and a single unprovoked seizure was hospitalized for severe thrombocytopenia (platelet 18,000/µL). Pseudothrombocytopenia was ruled out. He had no bleeding events, fever or sicca symptoms. No history of recent infection, immunizations, or introduction of new medications. Thoracoabdominopelvic CT scan showed bilateral axillary, mediastinal, and inguinal adenopathies. Biopsy of the axillary lymph node identified lipogranulomatous lymphadenitis lesions. Serologies for human immunodeficiency virus and hepatitis C antibody were negative, non-immune to hepatitis B virus. He had had previous contact with cytomegalovirus, Epstein-Barr and toxoplasmosis. Autoimmune study: anti-nuclear antibodies titer 1/1000, homogeneous pattern, anti-dsDNA 191 IU/mL, and positivity for anti-cardiolipin antibodies, beta2-glycoprotein, and lupus anticoagulant, increased complement consumption and circulating immune complexes. Diagnosis of SLE with secondary antiphospholipid syndrome was established, according to the 2019 criteria proposed by the European League Against Rheumatism and American College of Rheumatology. Treatment was initiated with prednisolone (1 mg/kg/day) and hydroxychloroquine (400 mg/day) with partial response (platelets 84,000/µL) after 22 days. Anticoagulation with warfarin was started after platelet counts consistently exceeded 30,000/µL.

Clinical manifestations of SLE are highly variable and can affect any system. Hematologic changes may include cytopenia, lymphadenopathy, and/or splenomegaly. Although rarer, since severe thrombocytopenia in SLE presents a very heterogeneous response to therapy, its rapid diagnosis and management are crucial.

Keywords: *thrombocytopenia, immune, systemic lupus erythematosus*

[Abstract:2184]

AUTOIMMUNE HEPATITIS IN A PATIENT WITH SYSTEMIC VASCULITIS

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Background: Autoimmune diseases tend sometimes to accumulate in the same individual. Autoimmune hepatitis and renal vasculitis have been occasionally reported with other autoimmune diseases, but the particular association of these disorders has rarely been previously reported in the medical literature.

Case Report: A 60-year-old female with a history of autoimmune thyroiditis and ANCA-negative systemic vasculitis was admitted to the department for abdominal pain and urticaria. The diagnosis of systemic vasculitis was made by part of the renal involvement with severe chronic renal failure, hematuria, proteinuria, and endocapillary and extracapsular glomerulonephritis at the renal needle biopsy. She was treated with corticosteroid and immunosuppressive treatment (Cyclophosphamide). Physical examination noted homogeneous hepatomegaly. There was no jaundice or splenomegaly. Hepatic tests revealed an important cytolysis. A liver biopsy showed characteristic features of autoimmune hepatitis (necrosis and inflammation). Serological tests were negative for viral hepatitis B and C. The diagnosis of autoimmune hepatitis (AIH) was retained following the detection of a high serum gammaglobulin level (over 1.5 times the upper normal limit), significant anti-nuclear antibodies level (over 1/320) and the result of liver biopsy with a Score of International Autoimmune Hepatitis Group (IAIHG) at 18 points. The patient was treated with corticosteroid and ursodesoxycholic acid. The patient developed acute pancreatitis and the evolution was fatal.

Conclusions: This observation is particular to the fact that renal vasculitis was associated with an AIH. This association is rare. The implicated mechanisms are unknown and lead to difficulties in the choice of suitable therapeutic management for this particular association.

Keywords: *hepatitis, systemic vasculitis, glomerulonephritis, autoimmune disease*

[Abstract:2185]

FEVER OF UNKNOWN ORIGIN, RELATING TO A CASE

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A 55-year-old heavy smoker, with no cardiovascular risk factors, presented with a two-month history of persistent fever, oral ulcers, and odynophagia. Despite two antibiotic cycles, the patient experienced no relief from the fever and lost 4-5 kg. Physical examination revealed mild anemia, leukocytosis, elevated ESR, and no other clinical abnormalities. A PET-CT scan indicated increased inflammatory activity in the thoracic and abdominal aorta, subclavian arteries, and brachiocephalic trunk, leading to a diagnosis of Takayasu's arteritis (TAK).

TAK is a rare large-vessel vasculitis, more prevalent in Asian women aged 10-40, with an unknown etiology. Clinical and radiological diagnosis involves constitutional symptoms, subacute fever, and subsequent vascular manifestations such as limb claudication, cyanosis, and hypertension. Laboratory tests, including elevated ESR and CRP, support the diagnosis. AngioMRI or angioCT is crucial for visualization of vascular changes, as biopsies are impractical.

Treatment involves high-dose systemic glucocorticoids, gradually tapered, supplemented with steroid-sparing agents like methotrexate or azathioprine. Anti-TNF-alpha or tocilizumab may be considered for maintaining remission. Endovascular interventions are reserved for irreversible stenosis or large

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aneurysms. This case underscores the importance of considering vasculitis in fever of unknown origin, particularly TAK, and highlights the role of advanced imaging techniques in diagnosis. The chronic and recurrent nature of TAK necessitates a comprehensive, individualized treatment approach to manage symptoms and prevent complications.

Keywords: *unknown fever, constitutional syndrome, Takayasu*



Figure 1. *CT-PET 1.*
Diffuse increase in inflammatory cellular activity throughout the entire length of the wall of the thoracic aorta and in the proximal two-thirds of the abdominal aorta up to its caudal portion to the renal hila. Increased metabolic activity in subclavian arteries and brachiocephalic trunk.

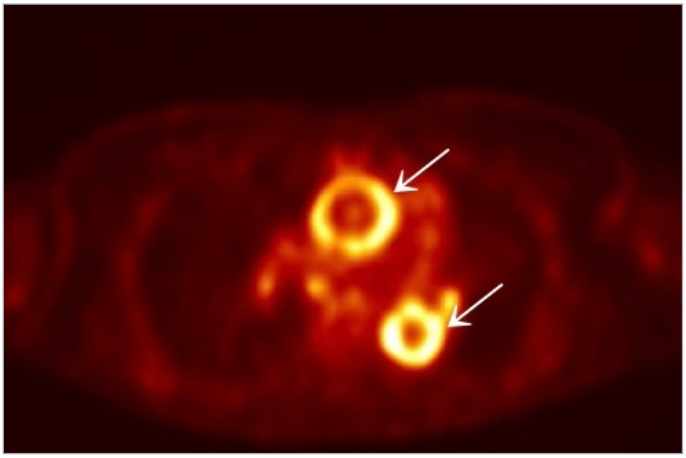


Figure 2. *CT-PET 2.*
Increase in inflammatory cellular activity throughout on the wall of the thoracic aorta.

[Abstract:2186]

ANTI-GLOMERULAR BASAL MEMBRANE (ANTI-GBM) DISEASE PRESENTING WITH PROLONGED CONSTITUTIONAL SYMPTOMS AND POSITIVE ANTI-MYELOPEROXIDASE (ANTI-MPO): A CASE REPORT

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Anti-glomerular basement membrane (Anti-GBM) disease is a type of small vessel vasculitis presenting with rapidly progressive kidney damage and alveolar hemorrhage. We present a case of anti-GBM disease presented with prolonged constitutional symptoms and anti-MPO antibody positivity.

Seventy-one-year-old female was referred to the rheumatology unit due to the preliminary diagnosis of interstitial lung disease and high acute phase. Despite having severe constitutional symptoms that lasted for more than 3 months, high acute phase responses and multiple hospital admissions, she could not receive any diagnosis. Although kidney function tests were normal two weeks ago, high creatinine and hyperkalemia were detected. The patient who had metabolic acidosis, hyperkalemia, oliguria and post-renal pathologies were ruled out, was urgently hospitalized and underwent hemodialysis. Pulse methylprednisolone, 1g IV, was started for 3 days for the rapidly progressive glomerulonephritis. On the second day of hospitalization, sudden hypoxia developed and alveolar hemorrhage was found. The patient was transferred to the intensive care unit because of the severe respiratory distress. Plasmapheresis with fresh frozen plasma, cyclophosphamide and IVIG was started. Then, Anti-GBM and anti-MPO were detected positive. The patient was lost following hypotension and bradycardia.

Several lines of evidence implicate that the rare distinct differences between patients with Anti-GBM and anti-MPO double-seropositive patients (DPPs) and single-positive patients. Compared with anti-GBM disease, DPPs tended to involve multi-organ damage rather than limited to the kidney. The disease may begin with obvious constitutional symptoms, and if diagnosis is delayed, it may have a fatal course due to rapidly progressive kidney and lung involvement.

Keywords: *anti-MPO antibody, Anti-glomerular basement membrane (Anti-GBM) disease, prolonged constitutional symptoms, interstitial lung disease, high acute phase*

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Figure 1. Thorax CT image of the patient: interstitial emphysema, pneumothorax on the right, subcutaneous emphysema and pneumomediastinum.

C reactive protein	360 mg/L	<5 mg/L
Sedimentation	122 Mm/h	0-25 Mm/h
Procalcitonin	1.15 ng/ml	
Leukocyte Count	13.8 K/uL	4.1-11.1 K/uL
Neutrophil count	9.1 K/uL	1.8-7.5 K/UL
Lymphocyte count	2.4 K/uL	1.2-5.8 K/uL
Eosinophil count	0.3 K/uL	0-0,5 K/uL
Hemoglobin	8.2 g/dL	12.6-17.8 g/dL
Platelet count	655 K/uL	150-400 K/uL
Ferritin	576ng/ml	
Blood Urea Nitrogen (BUN)	57 mg/dL	10-20 mg/dL
Serum creatinine	6.7 mg/dl	0.6-1.1 mg/dL
Alanine aminotransferase Aspartate aminotransferase	16 U/L 13 U/L	<33 U/L <32 U/L
Alkaline phosphatase Gammaglutamyl transferase	162 U/L 128 U/L	35-105 U/L 5-36 U/L
Lactate Dehydrogenase	173 U/L	135-225 U/L
Rheumatoid Factor	16.6 U/mL	<14 U/mL
Complement C3 Complement C4	1.5 g/L 0.28 g/L	0.9-1.8g/L 0.1-0.4g/L
ANA, IFA	1/80 Nucleoler patern	
Renal basement membrane antibodies	Glomerular +/tubular +	
Anti MPO	16.89 RU/mL	
Anti PR3	<2 RU/mL	
Anti nRNP/Sm Anti Sm	Negative	
Anti SS A Anti Ro 52	Negative	
Anti SS B Anti Scl 70	Negative	
Anti PM Scl Anti Jo 1	Negative	
Anti PCNA Anti Centromer B	Negative	
Anti Nukleozom Anti Histon	Negative	
Anti Ribosomal Protein Anti M2	Negative	
Anti DFS70	Negative	

Table 1. Results of the patient.

[Abstract:2188]

GASTRIC BANDING AND SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: The relationship between the implants of silicone and the appearance of autoimmune diseases particularly connective tissue disorders was reported in several publications. The responsibility of silicone in the pathogenesis of these connective tissue disorders by activation of the immune system does not make the unanimity of all authors.

We report a case of systemic lupus erythematosus (SLE) following the implementation of gastric banding in silicone.

Case Report: A 26-year-old female patient with a history of gastric banding 10 months ago presented with polyarthralgia and basic-thoracic pain. Physical examination revealed left pleural syndrome and joint pain with no fever. The diagnosis of pulmonary embolism was evoked in front of hypoxia, hypocapnia, and high levels of D-dimer, and confirmed by a thoracic CT scan. The diagnosis of SLE associated with the anti-phospholipid syndrome was retained in front of polyarthralgia, leuko-lymphopenia, positive antinuclear antibodies, positive native anti-DNA, positive anti-cardiolipins, and anti-Beta-2GPI antibodies. The evolution, under anticoagulants, was marked by the appearance of fever, right basis thoracic pain, dyspnea, and abdominal pain in front of gastric banding. An extension of the pulmonary embolism was confirmed. On the other hand, the barium swallow with gastro-duodenal follow-up showed indirect signs of banding migration. The patient was transferred to the intensive care unit.

Conclusions: Physio-pathological links between implants of silicone and connective tissue disorders remain very controversial. The relatively short delay of 7 months, between the implementation of gastric ring and the appearance of SLE would be in favour of the fortuitous character of this association?

Keywords: silicone, gastric banding, systemic lupus erythematosus, iatrogenic

[Abstract:2203]

EVALUATION OF THROMBOPHILIA RISK FACTORS IN BEHÇET'S PATIENTS WITH VASCULAR INVOLVEMENT

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Aim: The aim was to evaluate the presence of acquired or hereditary thrombophilic risk factors and effect of the presence

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of thrombophilia risk factors on vascular relapse development in Behçet's disease (BD) with thrombotic vascular involvement.

Materials and Methods: 225 patients with vascular involvement diagnosed with BD according to the International Behçet Study Group criteria were included. The presence of acquired and hereditary risk factors predisposing to thrombosis during vascular events was assessed.

Results: 141 patients were assessed for complete hereditary thrombophilia panel. The rate of patients with at least one acquired or hereditary risk factor in this group was 42.5%. The rate of patients with at least one hereditary risk factor was 28.4%, the rate of patients with at least one acquired risk factor was 23.4%. 46% patients were in the relapse group and 54% patients were in the non-relapse patient group. The presence of hereditary or acquired risk factors was found to be similar between the relapse and non-relapse groups. Although no statistically significant difference was detected, the proportion of patients without risk factors in the non-relapse group; the rate of patients with two or more risk factors and the total number of risk factors were found to be higher in the relapse group.

Conclusions: In BD with thrombotic vascular involvement, the hereditary risk factors presence during thrombotic events were found more frequent than in the general population, however it was lower than in venous thromboembolisms of general population. The presence of thrombophilic risk factors during the thrombosis in vascular BD had no effect on relapses.

Keywords: *vascular Behçet's disease, relapse, thrombophilia risk factor*

[Abstract:2209]

CHARACTERISTICS OF METABOLIC DISORDERS IN RHEUMATOID ARTHRITIS

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Objective: to determine the frequency of metabolic syndrome (MS) and its components in patients with rheumatoid arthritis (RA).

Materials and Methods: 29 patients with RA were included, divided into two groups: the first one (n=14) composed of patients with early (<2 years) clinical stages of RA and the second one (n=15) with late (>2 years) clinical stages of RA.

Results: MS occurred in 20 patients (69%), abdominal obesity in 19 (65.5%), hypertension in 18 (62%), hypercholesterolemia in 11 (37.9%), hyperuricemia in 11 (37.9%), hyperglycemia in 11 (37.9%), hypertriglyceridemia in 13 (44.8%) of 29 patients with RA. Depending on the presence of MS, patients were divided into 2 groups: I with MS (n= 20); II without MS (n=9). The duration of illness in patients with RA and MS was shorter, and disease activity according to the DAS28 and CDAI indices was higher than in patients without MS: 7.5 [6.5; 10.5] versus 12.0 [11.0; 22.0] months; DAS28 - 5.95 [4.9; 7.0] versus 4.1 [3.9; 4.2] points, CDAI - 34.8 [21.8; 41.4] versus 24.2 [18; 31] points accordingly ($p<0.05$ in all cases). MS components occurred with equal frequency in early and late stages of RA. In the group of patients with early RA, there was a correlation between the SDAI index (r=-0.34), BMI (r=-0.41), HDL cholesterol level (r=0.33), ESR (r=-0.35) and uric acid level (r=0.38).

Conclusions: In patients with early RA, a high frequency of MS and high disease activity determine a high risk of developing cardiovascular complications already at the onset of the disease.

Keywords: *metabolic syndrome, rheumatoid arthritis, cardiovascular events, hyperuricemia*

[Abstract:2249]

MORVAN SYNDROME SECONDARY TO THYMIC CARCINOMA IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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A 43-year-old female patient was admitted to the rheumatology clinic with complaints of fatigue, weakness, sleep disorders, and muscle fasciculations. In her history, she has been diagnosed with systemic lupus erythematosus and has carried the MEFV gene mutation since 2003. In thorax CT and PET/CT images, there was a 4x3.5 cm mass (Figure 1) in the anterior mediastinum suggestive of thymic carcinoma, FDG involvement accompanied by approximately 1 cm pleural thickening on the right anterior costal pleural surfaces, FDG accompanied by 5 mm pleural thickening on the costal-pleural surface on the right 5th rib involvement was detected. The patient underwent a thymectomy and excision of two pleural nodules. The pathological examination performed as a result of thymectomy was found to be compatible with thymic carcinoma. The surgical margin was found to be closer than 0.1 cm to the anterior mediastinal and posterior mediastinal margins. The patient was given 5 cycles of paclitaxel-carboplatin treatment. After the 4th cycle, a decrease in muscle fasciculations was detected. Bone marrow aspiration was found to be compatible with pure red cell aplasia due to thymic carcinoma. Follow-up of our patient continues in rheumatology, hematology, oncology, and neurology outpatient clinics. This case is being followed in our clinic as one of the rare cases where systemic lupus erythematosus Morvan syndrome and thymic carcinoma coexist.

Keywords: *morvan syndrome, thymic carcinoma, systemic lupus erythematosus*

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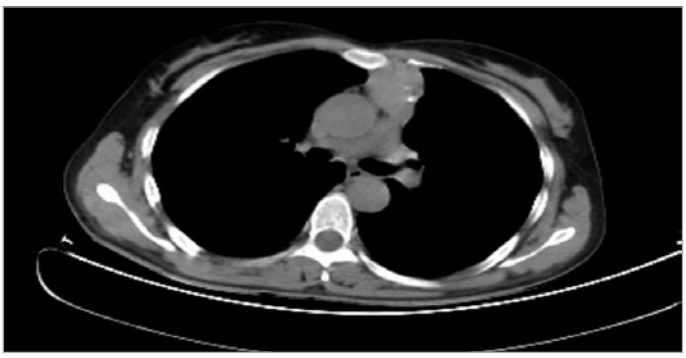


Figure 1. Thymic Carcinoma.

[Abstract:2257]

INTRAVENOUS CYCLOPHOSPHAMIDE AND METHYLPREDNISOLONE FOR INTERSTITIAL LUNG DISEASE WITH AUTOIMMUNE FEATURES (IPAF)

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Purpose: To describe the efficacy and safety of the regime consisting on cyclophosphamide-methylprednisolone (CYC-MP) as induction therapy for interstitial lung disease with autoimmune features.

Methods: Observational study of routine clinical care data of patients with IPAF according to the 2015 classification criteria, diagnosed between 2016 and 2022 and treated with a combination of intravenous CYC-MP followed by a reduced prednisone plus immunosuppressive regime, with a minimum follow-up of 12 months.

Findings: Eleven patients were included (54.5% women, mean age at diagnosis 69.6 years). 72.7% had non-specific ANA positivity, 18% anti-Ro positivity and 10% negative autoantibodies. All patients exhibited a radiological lung pattern by CT scan of Non-Specific Interstitial Pneumonia. All patients received induction therapy with intravenous CYC-MP, followed by maintenance therapy with reduced doses of prednisone combined with immunosuppressants (82% mycophenolate mofetil). The mean maximum prednisone dose was 7.7 mg/day (ranging from 15mg/d to 5mg/d). At the end of the follow-up time (range 12 to 64 months), the CT scan pattern improved or remained stable in 63.7% patients. The FVC improved in 36% and remained stable in 54%. The DLCO improved or remained stable in 73%. During 24 months of follow-up only 36.3% presented infectious complications, all mild. There were no dead to follow-up during the study.

Conclusions: Induction treatment with intravenous CYC-MP followed by maintenance therapy with reduced doses of prednisone and immunosuppressants either stabilized or improved radiological and functional parameters in more than 2/3 of patients with a low exposure to oral glucocorticoids and a good safety profile.

Keywords: *interstitial lung disease, IPAF, cyclophosphamide.*

[Abstract:2264]

IGG4-RELATED DISEASE: EXPERIENCE REGARDING NEW DIAGNOSTIC CRITERIA IN A FIVE CASE SERIES

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[2] Radiology Department
[3] Anatomic Pathology Department

IgG4-related disease is a systemic disease that have been recently grouped into four different phenotypes thanks to new diagnostic criteria that favour a better understanding of the complex diagnosis of this disease.

We present five cases observed in our Internal Medicine service in the last four years, reassessed with the diagnostic criteria proposed in 2019 by the American College of Rheumatology (ACR) and the European Alliance of Associations for Rheumatology (EULAR).

The presentation symptoms are often nonspecific, with a subtle progression leading to an average delay in diagnosis of two years. There is no clear gender predominance. Therefore, a comprehensive review of medical history, physical examination, and imaging tests is recommended for individuals with organ-limited involvement and elevated IgG4 levels.

It is not usual or essential to make a diagnosis of this disease based on the serological and histopathological studies of the lesions. A good response to corticosteroid treatment stands out in all cases. Biopsy is not essential for the diagnosis.

These criteria defined by researchers have been created considering only more commonly affected organs improving their specificity at the expense of a lower sensitivity, so extrapolating their results to our usual clinical practice should be done with caution.

Keywords: *IgG4-related disease, Inmunoglobulin G4, lymphoplasmocityc infiltrate, fibrosis, corticosteroids*

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	Case n°1	Case n°2	Case n°3	Case n°4	Case n°5
Age/Sex	47-year-old woman	64-year-old man	85-year-old woman	63-year-old man	60-year-old man
Symptoms	Neck pain, Abdominal and lower back pain	Abdominal pain	Fever and lower back pain	Lower back pain	Asymptomatic
Serum concentration of IgG4	Normal	Normal	Not measured	144 mg/dL	Normal
Biopsy	Yes	Yes	Yes	No	Yes
Score according to ACR/EULAR 2019 criteria	21	21	20	23	13
Diagnosis	Riedel's thyroiditis/Retroperitoneal fibrosis	Sclerosing mesenteritis	Periaortitis/Retroperitoneal fibrosis	Retroperitoneal fibrosis	Inguinal lymphadenopathy secondary to IgG4-related disease
Treatment/ Follow-up	Prednisone 30mg (2 years) + ramoxifen (3 years). Then azatioprine alone (3 years). Clinical remission. Radiological persistence of retroperitoneal fibrosis. Resolution of thyroid fibrosis at 3 years of follow-up.	Prednisone 30mg + MTX. Last 2 years prednisone 10mg. Clinical and radiological remission of lesions at 6 months on PET-CT, although uptake is detected in the right hip	Prednisone 50mg 7.5mg per day for one year Clinical remission at 2 years of follow-up. Persistence of retroperitoneal fibrosis at present	Prednisone 60mg 7.5 mg per day for one year Clinical remission during one-year follow-up. Persistence of retroperitoneal fibrosis at present.	Prednisone 40mg for 1 month, followed by 7.5mg per day for 6 months. No clinical manifestations reported during 6 months of follow-up

Table 1. *Summary of the clinical, analytical variables, and therapeutic response of the reported cases.*
MTX: Methotrexate Normal concentration of serum IgG4: 1.10-104 mg/dL PET-CT: Positron Emission Tomography-Computed Tomography

[Abstract:2291]

DOCTOR, MY KNEES HURT

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A 52-year-old man presented with arthralgias and fever for the last two weeks, with difficulty in ambulation. Furthermore, he associated oppressive chest pain and dyspnea with moderate exertion. He was a professional plumber and was a non-dependent person. His personal history included active smoking, arterial hypertension, chronic obstructive pulmonary disease and a transient cerebrovascular accident. On arrival, the patient was stable, with 38°C temperature. The physical examination was unremarkable. Laboratory tests, electrocardiogram, X-rays and blood cultures were requested. Leukocytosis was 19800, PCR206 and troponin I1131. He was evaluated by Cardiology Service who performed an echocardiogram that was normal, with a diagnosis of stable angina with acute myocardial damage related to an inflammatory or infectious process. He was admitted to the Internal Medicine Service with suspicion of autoimmune/inflammatory pathology as the first possible diagnosis. Temperature maintained at 39°C. An autoimmune study, serology and blood cultures with fever was requested and symptomatic treatment started. The positivity for AC anti-proteinase 3 and anti-citrullinated peptide stood out, as well as positive rheumatoid factor and elevated ESR. A catheterization was requested, which was normal, and 24-hour urine was found to have mild proteinuria in the non-nephrotic range. Oral prednisone was started and he was evaluated by Rheumatology Service, with a diagnosis of seropositive rheumatoid arthritis and ANCA-positive vasculitis.
He was discharged from the hospital with oral prednisone 15 mg daily with a downward pattern and is scheduled for outpatient consultations, where he was started treatment on rituximab and methotrexate.

Keywords: *vasculitis, prednisone*

[Abstract:2292]

CLINICAL FEATURES OF MRI VERIFIED CHANGES IN THE ATLANTOAXIAL REGION IN PATIENTS WITH ANKYLOSING SPONDYLITIS

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The inflammatory process in patients with ankylosing spondylitis (AS) can cause changes in the atlantoaxial region (AAR) with atlantoaxial instability (AAI), which potentially leads to various clinical manifestations and irreversible neurological complications.
Purpose: to evaluate the clinical features of AS in patients with MRI verified changes in the atlantoaxial region.
Materials and Methods: 30 patients with AS underwent neck MRI. The Average age was 46.0±13.8. Activity (ASDAS(CRP) was very high for 83.4%, high for 10%, low for 6.6%. The median duration of illness 190.5 [2-408] months. Comorbid pathology was 66.7%, median 3 [0-13]. Clinical examination included the assessment of neuropathic pain (NP) according to the Pain detect (PD), central sensitization (CS). CSI neurological status was assessed. AAI was assessed using MRI of the neck with measurements of craniometric parameters, and then deviations were calculated in comparison with MRI data of 45 healthy individuals.
Results: 76.7% had neck pain, 96.7% inflammatory, 80% non-inflammatory pain (CS–53.3%, NP-70%). The average pain value according to VAS was 6.1±2.2. MRI changes in the AAR were 60%. Neck pain was associated with NP (rSp=0.378;p=0.039), pain intensity was higher in patients with CS (rSp=0.611;p=0.001). There was a relationship between AAI and the duration of the disease (rSp=0.499;p=0.005), activity (rSp=0.398;p=0.032), number of tender (rSp=0.505;p=0.009) and swollen joints (rSp=0.471;p=0.015), number of comorbid diseases (rSp=0.471;p=0.009).
Conclusions: Most patients with AS have initial AAR changes on MRI. The pain is of great intensity, mixed in nature with a predominance of the neuropathic component. AS activity, duration and number of comorbid diseases are higher in the presence of such changes.

Keywords: *ankylosing spondylitis, atlantoaxial region, atlantoaxial instability*

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[Abstract:2303]

UNRAVELING RHABDOMYOLYSIS: NOT ALWAYS IS IT EXTREME EXERCISE

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Inflammatory myopathies are autoimmune diseases that affect multiple organs and systems, characterized by muscular weakness and inflammation.

A 30-year-old man, without relevant medical history, presented with muscle pain, weakness, and dark-colored urine. In the emergency department, significantly elevated levels of creatine phosphokinase (CPK) were observed, with peak levels reaching 271,500 IU/L, lactate dehydrogenase (LDH) at 7,600 IU/L, aspartate aminotransferase (AST) at 1,303 U/L, and alanine aminotransferase (ALT) at 183 U/L. As a result, he was admitted to the intensive care unit and discharged upon analytical improvement with a diagnosis of inflammatory myopathy, specifically antisynthetase syndrome.

Serological tests revealed negative viral serologies, positive antinuclear antibodies (1/80 titer, dense fine speckled pattern AC19), and presence of antisynthetase antibodies (anti-OJ, anti-PL12, and anti-PL7). Electromyography (EMG) and nuclear magnetic resonance imaging (MRI) suggested an acute myopathic process. Rhabdomyolysis, although more commonly caused by trauma, drugs, or infections, can also result from autoimmune myopathies, falling under idiopathic inflammatory myopathies, including various entities such as dermatomyositis, polymyositis, and autoimmune necrotizing myositis.

The treatment involved intravenous fluid therapy to prevent renal damage, followed by three days of steroid boluses and subsequent tapering to a dose of 1 mg/kg of body weight. The patient showed good progress, remaining asymptomatic with a decreasing steroid regimen.

Rhabdomyolysis due to autoimmune myopathies is uncommon, but its recognition is crucial for proper management. Specific tests, such as antisynthetase antibodies, along with imaging studies and muscle biopsy, are essential for diagnosis. Additionally, treatment varies based on etiology, with immunosuppressants required in autoimmune cases.

Keywords: *creatine phosphokinase (CPK), rhabdomyolysis, autoimmune myositis, electromyography (EMG)*

[Abstract:2316]

IMPACT ON PATIENT OUTCOME WITH RENAL INVOLVEMENT IN PRIMARY SJOGREN'S SYNDROME: A MORE SERIOUS COMPLICATION THAN PREVIOUSLY THOUGHT

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Summary: Renal disease in primary Sjogren's syndrome (pSS) is associated with increased rates of chronic kidney disease (CKD), hospitalisation and mortality.

Purpose: The aim of this study is to investigate the progression to chronic kidney disease or end-stage renal disease in patients diagnosed with primary Sjogren's syndrome with renal involvement and its impact on mortality at a tertiary hospital.

Methods: A retrospective analysis of pSS patients diagnosed between 1993 and 2023 at a single centre was conducted. Descriptive analysis was performed on the evolution of renal deterioration. Bivariate and multivariate analysis was performed to examine the influence of renal disease on patient outcome.

Findings: The study included 252 patients, 56 (22.2%) had renal involvement. In patients with renal involvement, 24.1% had progressive deterioration of renal function and 1.85% developed end-stage renal disease. The bivariate analysis indicated a trend towards a higher mortality rate within this subgroup, although it was not statistically significant (16.1% vs 7.14%, p=0.074). There was a statistically significant rise in complications (42.9% vs 17.9%, p<0.001), particularly of the infectious kind (23.2% vs 7.73%, p=0.003). However, there was no evidence that renal impairment increased mortality (OR 1.45, 95% CI 0.49 to 4.02, p=0.486), which may be due to small sample size.

Conclusions: Our study highlights the importance of closely monitoring patients with pSS for signs of renal involvement. Early identification and treatment of kidney disease may prevent decline in renal function and development of serious complications.

Keywords: *Sjogren's, renal disease, prognosis*

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[Abstract:2323]

IMMUNOSUPPRESSIVE TREATMENT IN PRIMARY SJOGREN'S SYNDROME WITH RENAL DISEASE

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Summary: Management of renal disease in primary Sjogren's syndrome (pSS) is based on the correction of hydroelectrolyte imbalances and immunosuppressive treatment. However, the evidence for the use of immunosuppressive therapy is limited.

Purpose: To assess the impact of immunosuppressive therapy on the progression of renal function decline in pSS patients with renal disease and its effect on mortality at a tertiary hospital.

Methods: A retrospective analysis of pSS patients with renal disease diagnosed between 1993 and 2023 was conducted. A bivariate analysis was performed on immunosuppressive treatment and its effect on the development of renal dysfunction and vital prognosis in these patients.

Findings: The study included 252 participants. Fifty-six (22.2%) had renal involvement, of whom 24.1% had worsening renal function. One patient progressed to end-stage renal disease (ESRD). Immunosuppressive treatment was given to 26.6% of patients, with systemic corticosteroids being the preferred treatment. Prednisone was used in 24.2% of patients, while 3.97% received methylprednisolone. Additional therapies included: methotrexate (6.75%), rituximab (5.95%), azathioprine (4.37%), mycophenolate mofetil (2.38%) and cyclophosphamide (0.79%). Bivariate analysis revealed that patients receiving methylprednisolone and azathioprine were significantly more likely to achieve complete remission or improvement in renal function than to experience worsening of renal function or progression to ESRD (83.3% vs 0%, p=0.039 for both outcomes). There was no statistically significant effect of immunosuppressive treatment on survival.

Conclusions: Our study shows the importance of immunosuppressive treatment in preventing the development of renal dysfunction and progression to ESRD, reinforcing the evidence for the use of systemic corticosteroids.

Keywords: *Sjogren's, renal disease, immunosuppressive treatment*

[Abstract:2339]

SYSTEMIC LUPUS ERYTHEMATOSUS DIAGNOSIS WITH GASTROINTESTINAL SYSTEM INVOLVEMENT

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Case Description: a 28 years old female patient with epilepsy and thrombocytopenia during 10 years applied to our emergency clinic with complaints of severe abdominal pain (especially after meals) and weight loss (from 46 kg to 39 kg in 4 weeks). She had been receiving carbamazepine-zonisamide treatment for epilepsy. Her brother had FMF, her father had colon Ca, her mother was under follow-up for thrombocytopenia. The general condition was poor due to abdominal pain, the patient could not stand upright or walk due to pain. TA: 82/51 mm/Hg, radial pulse: 92/min, heat: 36.8°C. She had cachectic appearence, BMI:16.7 kg/m^2.

Bilateral neck, axillary, inguinal lymph nodes were palpated. During the abdominal palpation, there was muscular defense; traube's area was closed. There was asymmetric polyarthritis in the wrists, ankles, elbows, proximal-distal interphalangeal joints. Sedim: 5, CRP: 37 mg/L (0-5), Hgb: 98 g/L (115-155), MCV: 74.4 fL (80-95), WBC: 2.13.103/uL (3.8-10), Neu :1.51.103/uL (1.56-6.13), lym: 0.38.103/uL (1.18-3.74), PLT: 85.103/uL (150-400.103), kre: 0.42 mg/dL (0.50-0.90), AST:27U/L (0-32), ALT: 49U/L(0-33),ALP: 224U/L (35-104), GGT: 336U/L (6-42), LDH: 207U/L(135-214), total protein: 58g/L (66-83), albumin: 4g/L (35-52). Abdomen USG was normal.

Clinical Hypothesis: Differential diagnosis was mesenteric arterial vasculitis, malignancies, infectious or autoimmune conditions.

Diagnostic Pathways: Contrast Enhanced Abdominal MRI, Abdominal CT, abdominal angiography showed mild hepatosplenomegaly. ANA: positive (1/1260, diffuse homogenous), anti-ds-DNA: >800 IU/ml positive (<100), ENA: positive, Coombs: positive (direct: 2+, indirect: 1+), C3: 0.63 g/L (0.9-1.8), C4: 0.07g/L (0.1-0.4). Lupus anticoagulant, anti-Cardiolypin-IgM-IgG, anti-Beta2 glycoprotein IgM-IgA-IgG; ASMA, anti-LKM, AMA were negative. Urine test was normal. ECG revealed negative T waves in inferolateral derivations, evaluated as pericarditis by cardiology consultation. Ecocardiography reported pericardial hyperechogenity and mild pericardial effusion on right atrium. Thorax CT was normal. Gastroscopy demonstrated no pathology. Bilateral neck, axillary, inguinal ultrasonography detected multiple lymph nodes (>2cm). 18-FDG-PET-CT demonstrates inflammation of cecum (SUVmax: 13.5). The patient was diagnosed with systemic lupus erythematosus, based on the available clinical-laboratory findings. Methylprednisolone, hydroxychloroquine; anticoagulant, antiaggregant treatments were initiated. The patient's complaints relieved from the first days of treatment. The patient is still under our follow-up in remission.

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Discussion and Learning Points: Small and medium vessels vasculitis at gastrointestinal system can be seen in SLE. Mesenteric vasculitis is rarely seen in SLE patients with multiple organ involvement and active disease.

Keywords: *systemic lupus erythematosus, gastrointestinal system, vasculitis*

[Abstract:2343]

THE RELATIONSHIP OF SYNOVITIS WITH THE RISK KNEE OSTEOARTHRITIS PROGRESSION

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Purpose: To study the relationship of synovitis with the risk of knee osteoarthritis (OA) progression with long-term follow-up.
Methods: The study included 110 women (59.11±8.95 y.) with knee OA (ACR), who were examined twice with an interval of 5 years. The mean duration of OA was 8 [3;20] years. All patients underwent a questionnaire including clinical data, assessment of knee pain on a visual analog scale, radiography and ultrasound (US) examination of the knee.
Results: After 5 years of follow-up, X-ray progression was detected in 40 patients (group 1), in 70 (group 2) - the stage of OA did not change. Patients in both groups were comparable in age (59.2±9.5 and 59.0±8.1 y.) and duration of the disease (11.1±10.6 and 13.7 ±9.9 y.), however, patients in group 1 had significantly higher body mass index (BMI) (34.18 ± 6.24 versus 31.56 ± 5.45, p = 0.024), more intense knee pain when walking (72.03 ± 19.32 vs. 58.44 ± 13.3, p = 0.001), synovitis was more often detected on US: 62.5 vs. 34.3% (p = 0.004). When conducting a correlation analysis (Spearman) between the X-ray stage and the factors of progression of knee OA, the following correlation coefficients were identified: knee pain when walking (r = 0.44, p<0.05), BMI (r = 0.32, p<0.05), knee synovitis by US (r = 0.49, p <0.05). Multivariate analysis confirmed that synovitis is a significant factor in knee OA progression (p<0.05).
Conclusions: Synovitis is one of the significant risk factors for progression in patients with knee OA.

Keywords: *osteoarthritis, synovitis, knee pain*

[Abstract:2353]

A CASE OF FAMILIAL MEDITERRANEAN FEVER WITH AMYLOID STORM DEVELOPING AFTER HEART VALVE REPLACEMENT

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Case Description: We report the case of a 39-year-old female admitted with diarrhea, vomiting, and decreased urine output. After aortic and mitral valve replacement surgery performed 3 weeks before, her complaints started and worsened. She had diagnosis of familial type 2A hyperlipidemia and FMF. She was under ezetimibe, acetylsalicylic acid, pantoprazole, bisoprolol, warfarin, atorvastatin, colchicine, canakinumab. On physical examination generalized and bilateral 3+ pretibial edema, ascites, serous discharge in the sternotomy area. Laboratory showed CRP 104 mg/dL, creatinine 3.6 mg/dL, proteinuria 30 gram/day and amyloid storm (AS) triggered by surgery and gastroenteritis was considered. Anakinra 200 mg/day was started and titrated according to clinical and laboratory findings up to 600 mg/ day. Both intermittent hemodialysis and regular lipid apheresis were continued. Ten days later, acute calculous cholecystitis developed and cholecystectomy was performed. After 2 weeks of ICU support she died due to septic shock.
Clinical Hypothesis: AS should be considered in acute, rapidly progressive renal failure, increased proteinuria and high acute phase reactants.
Diagnostic Pathways: Abrupt renal and inflammatory findings in FMF patients suggests AS.
Discussion and Learning Points: AS is a rare complication of FMF disease that can develop even in the absence of systemic amyloidosis and renal dysfunction, but is a complication of FMF disease that has the highest morbidity and mortality in the acute phase. Although it is known that it is often triggered by infection, it should be kept in mind that other stressors can also cause amyloid storm. Early recognition of this condition and rapid initiation of anti-IL1 treatments makes a significant prognostic difference.

Keywords: *amyloid, serositis, hemodialysis*

[Abstract:2367]

CARDIAC INVOLVEMENT IN INFLAMMATORY MYOPATHIES

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Introduction: Inflammatory myopathies (IM) are subgroups of autoimmune diseases characterized by muscle inflammation.

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Cardiac involvement in IM is associated with bad prognosis. The aim of this study is to determine the clinical and paraclinical characteristics and the prognostic impact of cardiac involvement in IM.

Patients and Methods: A retrospective descriptive monocentric study conducted on the medical records of patients with IM who developed cardiac involvement related to the disease over a period of 17 years (2005-2022).

Results: A total of 97 patients were collected, among whom cardiac involvement was found in 14 patients (14%), including eight men and six women (gender ratio of 1.3). The average onset time of cardiac involvement after the initial diagnosis of IM was 12.35 months. It was symptomatic in ten patients (71%). Functional signs included dyspnea (n=8) and chest pain (n=6). Six patients had rhythm disturbances on electrocardiogram. Pericardial involvement was found in nine patients. Myocarditis was observed in eight patients. Specific cutaneous signs were present in all patients. The identified IM included dermatomyositis (n=2), anti-synthetase syndrome (n=6) (including four with anti-Jo1 antibodies, one patient with anti-PL12 antibodies, and another with anti-PL7 antibodies), inclusion body myositis (n=1), and autoimmune necrotizing myositis with anti-SRP antibodies (n=2). All patients were treated with cyclophosphamide combined with corticosteroids at a dose of 1mg/kg/day. The outcome was favorable in all patients.

Conclusions: Cardiac involvement represents a serious condition in IM that can jeopardize patients' lives. Its investigation should be systematic due to its relatively frequent subclinical nature.

Keywords: *inflammatory myopathy, cardiac involvement, prognosis*

[Abstract:2370]

ANTIPHOSPHOLIPID SYNDROME ANDSWJOGREN'S SYNDROME WITH AUTOIMMUNE ADRENALITIS

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Case Description: A 23-year-old female with Hashimoto's thyroiditis presented with acute double vision. Cranial imaging revealed dural sinus thrombosis. Thrombocytopenia and elevated amylase, lipase, and acute phase reactants were shown in laboratory. Abdominal imaging was performed due to elevated amylase and lipase and abdominal pain, confirming acute pancreatitis. Furthermore, bilateral adrenal hemorrhage was detected. She was exhibited high ACTH and low cortisol levels and replacement with hydrocortisone was given. To investigate autoimmune features; rheumatologic serology, thrombophilia mutations, paroxysmal nocturnal hemoglobinuria clone and other hematological examinations were requested. Only positivity was found on triple antiphospholipid (lupus anticoaguant, anti-cardioplipin IgM and IgG, anti-beta 2 glycoprotein IgM and IgG), anti-Ro and anti-La antibodies.

The Schirmer test showed significant bilateral dry eyes, and the salivary gland biopsy supported Sjogren's syndrome (Focus score > 1). The patient was referred to Rheumatology for follow-up after being diagnosed with autoimmune adrenalitis induced by antiphospholipid syndrome + Sjogren's syndrome. Under corticosteroids, antiaggregants, anticoagulants and mycophenolate mophetil combination, all symptoms and laboratory abnormalities resolved.

Clinical Hypothesis: When evaluated in conjunction with the diagnosed thrombosis and accompanying laboratory findings, autoimmune and hematological diseases were predominantly considered.

Diagnostic Pathways: In case of one autoimmune feature and unexplained systemic finding, other autoimmune disorders should be investigated.

Discussion and Learning Points: A multidisciplinary approach is crucial in patients systemic findings, thrombosis and autoimmunity. Adrenalitis and adrenal hemorrhage are so rare presentations and concomitant thrombosis should lead us to screen for autoimmune conditions in young female patients.

Keywords: *thrombosis, autoimmunity, adrenal hemorrhage, Sjogren's syndrome, antiphospholipid syndrome*

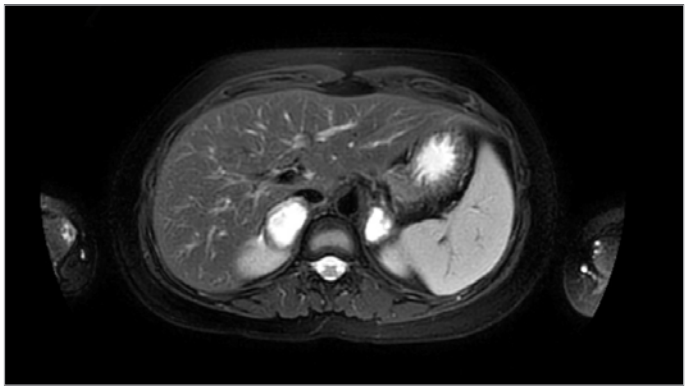


Figure 1. *MRI.*
Bilateral adrenal hemorrhage.

[Abstract:2375]

TOCILIZUMAB: A PROMISING REMISSION TREATMENT IN TAKAYASU ARTERITIS

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Introduction: Takayasu arteritis (TA) is a vasculitis affecting large-caliber vessels, especially the aorta and its main branches. Tocilizumab (TCZ) treatment (humanized monoclonal antibody against the IL-6 receptor) has been proposed, particularly in

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patients with refractory TA. We report a case of refractory TA effectively treated with tocilizumab.

Case: A 48-year-old female with TA since the age of 33. Aortic angio-scan showed circumferential and regular thickening of the ascending, and descending thoracic aorta, and the right renal artery. There was sub-occlusive stenosis of the left subclavian artery and tight stenosis of the common carotids. Initially treated with three perfusions of Solumedrol, followed by oral corticosteroids (1 mg/kg/day) and methotrexate (MTX) (10 mg/week), along with antihypertensive treatment, leading to significant improvement of initial symptoms.

The patient experienced a first relapse in 2020 with recurrence of claudication in the upper and lower limbs, syncopal episodes, and radiological worsening. Treated with IV and oral corticosteroids and an increase in MTX dose to 15 mg/week. She underwent dilatation of the common carotid artery and post-dilation stenting of the residual stenosis with good angiographic control. A third flare in 2022 worsened the initial lesions and new stenosis occurred on previously healthy arteries (both vertebral arteries), requiring an increase in corticosteroid and MTX and endovascular treatment with no improvement. Tocilizumab treatment was initiated. The patient received 14 courses (8 mg/kg/month) with favourable clinical and radiological outcome

Conclusions: Our case demonstrates the efficacy of tocilizumab in the treatment of refractory and corticosteroid-dependent TA.

Keywords: *Takayasu arteritis, tocilizumab, refractory*

[Abstract:2386]

DOUBLE ANAEMIA - A RARE CASE OF PERNICIOUS ANAEMIA CONCOMITANT WITH AUTOIMMUNE HEMOLYTIC ANAEMIA

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Pernicious anaemia is a megaloblastic anaemia caused by a deficiency of B12 vitamin induced by autoantibodies that prevent its absorption and cause autoimmune atrophic gastritis. In autoimmune hemolytic anaemia, erythrocytes are destroyed by autoantibodies, which can act at body temperature ("hot" antibodies) or below it, usually in the extremities ("cold" antibodies). The occurrence of these two entities simultaneously is rare.

We present the case of a 51-year-old woman with a two week history of asthenia and anorexia. Blood tests revealed pancytopenia, indirect hyperbilirubinemia, significant increase in lactate dehydrogenase, and greatly decreased haptoglobin levels. Peripheral blood smear showed abundant erythrocyte agglutination with improvement after heating, macro ovalocytes, teardrop cells, and hypersegmented neutrophils. Urinary sediment was normal. She was admitted with a diagnosis of autoimmune hemolytic anaemia and put on prednisolone 1 mg/kg/day. Complementary investigation revealed a strong positive Coombs test for IgG and C3d and the presence of anti-I antibody (cryoagglutinin) at a titer of 1:64. Deficiency of B12 vitamin <125 pg/ml and positive anti-parietal cell and anti-intrinsic factor autoantibodies were also detected, confirming the simultaneous diagnosis of pernicious anaemia.

In the presence of megaloblastic anaemia with hemolysis and positive Coombs test, a careful and systematic approach is necessary. We know that occasionally, pernicious anaemia may be the cause of hemolysis, and in rare cases may be accompanied by a positive direct Coombs test. It is also associated with higher incidence of other autoimmune diseases. Here, the presence of cryoagglutinins suggests the rare coexistence of autoimmune hemolytic anaemia and pernicious anaemia.

Keywords: *pernicious anemia, autoimmune hemolytic anemia, cryoagglutinin, cold agglutinin*

[Abstract:2393]

INFLAMMATORY MYOPATHY IN MEN

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Introduction: Inflammatory myopathies (IM) are rare autoimmune diseases characterized by a heterogeneous clinical presentation dominated by muscular involvement. The aim of our study was to determine the clinical and paraclinical characteristics of IM in men.

Patients and Methods: A retrospective descriptive monocentric study was conducted on the medical records of men with inflammatory myopathies over a period of seventeen years (2005-2022).

Results: Among 97 patients with inflammatory myopathies, 27 were men. The average age at the time of diagnosis was 56 years. Myalgia was present in 70% of cases. General symptoms were observed in four patients. Proximal muscle weakness was present in 88% of cases. Five patients had dysphonia related to palatal muscle paralysis. Two patients experienced swallowing difficulties. Joint involvement was reported in nine patients (33%). Diffuse infiltrative pneumopathy was observed in ten patients (38%), three of whom were asymptomatic. Myocarditis was observed in 25% of cases. Creatine phosphokinase (CPK) levels were elevated in all patients. Electromyography showed a myogenic pattern in 21 patients. Antisynthetase syndrome was present in nine patients. One patient had dermatomyositis with anti-MDA5 antibodies. Two patients had positive anti-SRP antibodies. One patient had inclusion body myositis. Three patients had neoplasms.

All patients received oral prednisone. Immunosuppressants, including methotrexate (14 cases) and cyclophosphamide (nine cases), were given to 23 patients. The outcome showed remission

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in 11 cases, stabilization in seven cases, and worsening in 12 cases.
Conclusions: Cardiac and pulmonary involvement, seem to be frequent in men with IM.

Keywords: *inflammatory myopathy, men, characteristics*

[Abstract:2398]

INSIGHTS INTO SEVERE HEMOPHAGOCYTIC SYNDROME: CLINICAL PROFILES, DIAGNOSTIC CHALLENGES, AND TREATMENT OUTCOMES IN A RETROSPECTIVE STUDY AT HOSPITAL DE SALAMANCA

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Introduction: Hemophagocytic Syndrome (SHF), a severe immunological disorder, exhibits high in-hospital mortality, marked by excessive immune activation and multiorgan failure triggered by drugs, infections, or predisposing genetic/acquired conditions. Analyzing 15 SHF diagnosed Internal Medicine patients, we reviewed primary causes, common clinical manifestations, laboratory findings, treatment and mortality.
Materials and Methods: A retrospective observational study on SHF was conducted in Internal Medicine department of the Hospital de Salamanca (2012-2023). Electronic health records provided biographical, clinical and diagnostic data for fifteen patients.
Results: Among 15 patients (average age: 61, 5 females), fever (93%) and headache (26%) were primary symptoms. Laboratory findings included elevated triglycerides (86%), LDH (93%), and ferritin (100%). Probable infectious etiology was identified in 9 cases, and 3 were autoinflammatory/autoimmune. Treatment comprised antibiotic therapy (66%), corticosteroids (53%), and other immunological drugs (26%). In-hospital mortality was 26%.
Conclusions: This study delves into SHF patients' clinical and analytical features, emphasizing the prominence of fever and specific laboratory abnormalities. Prevalent infectious and autoimmune etiologies were observed. Treatment involved antibiotics and corticosteroids, with a 26% mortality rate.
Swift management and identifying underlying causes are crucial. Diagnostic and prognostic markers, like the HScore by Laurence et al., are emphasized, underscoring SHF severity and the need for more effective diagnostic and therapeutic strategies.

Keywords: *hemophagocytic syndrome, hemophagocytic lymphohistiocytosis, internal medicine, Salamanca*

[Abstract:2400]

THERAPEUTIC CHALLENGE IN ANTIPHOSPHOLIPID SYNDROME WITH LONG-SANTING NEUROLOGICAL FOCALITY

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Purpose: Long-standing neurological focality without evidence of ischemic or hemorrhagic pathology on imaging tests presents a diagnostic challenge given the wide range of possible differential diagnoses. Among them we find antiphospholipid syndrome, which can present as primary pathology or secondary to other autoimmune diseases, most frequently systemic lupus erythematosus. Once the diagnosis has been made, we should consider adequate thrombotic prevention, both primary and secondary, given the high morbimortality of this disease.
Case Report: In the present work we studied the case of a 60-year-old woman with dysarthria. The patient referred alternating neurological clinic of two years of evolution under study by neurology.
Findings: After performing cranial CT, cranial Angio-CT, TTE and TSA Doppler without pathological findings the patient was discharged pending cranial MRI with subsequent control in consultations. One month later the patient was summoned to report lesions compatible with demyelinating disease accompanied by triple positivity in antiphospholipid antibodies.
Conclusions: This case represents the need for extensive studies for neurological focality, not only from a neurological point of view. It also poses a therapeutic challenge as it does not fit into primary or secondary prevention protocols and the importance of individualizing each treatment.

Keywords: *antiphospholipid syndrome, antiphospholipid antibodies, demyelinating lesions.*

[Abstract:2405]

INFLAMMATORY MYOPATHIES (IM) WITH LATE ONSET IN THE ELDERLY

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Introduction: Muscular disorders in the elderly are mainly dominated by secondary myopathies. Inflammatory primary myopathies (MII) are rarer, often progress slowly, and are frequently associated with neoplasms.
The objective of our study was to describe the clinical, laboratory, and therapeutic characteristics of MII in the elderly.
Patients and Methods: This is a descriptive, retrospective, and monocentric study conducted on the records of patients diagnosed with MII over a period of 17 years (2005-2022).

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Results: Among the 97 patients monitored for MII, eighteen (17%) were elderly at the time of diagnosis, with five being over 70 years. The average age was 67 years with a male-to-female ratio of 0.8. General symptoms were present in seven patients (38%). Myalgias were reported in ten patients. Muscular belt weakness was present in all patients. No patient had difficulty swallowing. Articular involvement was reported in 57.1% of cases. Diffuse infiltrative pneumopathy was observed in ten patients (58%). Cardiac involvement was observed in 14 cases (20%). The average elevation of creatine phosphokinase (CPK) was 6.5 times the normal level. Muscle biopsy was performed in seven patients, revealing pathological findings in five cases. Antisynthetase syndrome was present in five patients. One patient had dermatomyositis. Six patients had neoplasms at the time of diagnosis: four colorectal adenocarcinomas, one endometrial adenocarcinoma, and one pulmonary adenocarcinoma.
Conclusions: The diagnosis of MII in the elderly remains challenging due to the frequent association with other diseases, but it must be made early to exclude an underlying neoplasm.

Keywords: *elderly, inflammatory myopathy, prognosis*

[Abstract:2415]

LOW BACK PAIN. ARE WE CARING ENOUGH?

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Case Description: A 53-year-old male referred to the Internal Medicine clinic due to bone lesions detected in the context of long-standing lower back pain. He did not report weight loss, asthenia, reduced appetite, fever, or any other symptoms indicative of a neoplastic or infectious origin. There were no notable abnormalities found in the physical examination or in the urgently conducted basic complementary tests (chest X-ray, blood tests, electrocardiogram).
Diagnostic Pathways: A body CT scan and MRI of the spine were performed for a more detailed characterization of the lesions. The findings revealed only intrathoracic reactive lymph nodes and bone lesions in the spine with a mixed lytic-blastic pattern. This initially suggested a neoplastic origin as the primary possibility. Given a normal PSA result, a bone biopsy guided by CT was performed. The histopathological report revealed non-caseating granulomas. This finding, along with intrathoracic lymphadenopathy and elevated angiotensin-converting enzyme, suggested the possibility of sarcoidosis with nodal and osseous involvement.
Discussion and Learning Points: Sarcoidosis is a condition characterized by highly diverse clinical manifestations. While the lungs are frequently affected, the granulomatous process can also extend to involve bones. Identifying and diagnosing sarcoidosis in bones is generally straightforward, but the treatment outcomes are often unsatisfactory. In our case, treatment was initiated with prednisone at a dose of 0.5 mg/24h, achieving an excellent clinical response. After one year of treatment, corticosteroid therapy has been successfully discontinued, and the patient has remained asymptomatic to date.

Keywords: *sarcoidosis, bone, low back pain*



Figure 1. *MRI.*

[Abstract:2416]

CRYOGLOBULINEMIC VASCULITIS AND ANCA-ASSOCIATED VASCULITIS: A GENUINE ASSOCIATION?

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Introduction: The coexistence of granulomatosis with polyangiitis (GPA) and cryoglobulinemic vasculitis is rare and poses both diagnostic and therapeutic challenges. We report the case of a patient with both cryoglobulinemic vasculitis and GPA.
Case Report: A 53-year-old female with no notable medical history presented with vascular purpura on the lower limbs. She reported ongoing lower limb paresthesia for two months and recurrent nasal obstruction and epistaxis. Cardiac and pulmonary auscultation was unremarkable, as was the rest of the examination. Laboratory tests showed elevated inflammatory markers and normal renal function. Blood cultures and viral serologies were negative. Cardiac ultrasound was normal. Lower limb electromyography indicated a length-dependent axono-demyelinating sensory-motor polyneuropathy. Thoraco-abdomino-pelvic CT scan revealed excavated pulmonary nodules. Skin biopsy showed features of pyoderma gangrenosum. Due to mediastinal adenopathy, a biopsy revealed giant cell granulomas. Bronchoalveolar lavage indicated a Gold score of 150, confirming

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intra-alveolar hemorrhage. Facial CT scan showed septal deviation, and nasal biopsy revealed leukocytoclastic vasculitis. Cryoglobulin testing was positive for IgG and IgA. Immunological assessment demonstrated positivity for anti-PR3 C-ANCA. Based on clinical and paraclinical evidence, the diagnosis of GPA associated with cryoglobulinemia was established. The patient was treated with 60 mg/day of corticosteroids, and cyclophosphamide perfusions. The clinical and radiological evolution was favorable.

Conclusions: This case emphasizes the need for a comprehensive evaluation in complex clinical situations, especially when multiple, infrequently associated diagnoses coexist in the same patient.

Keywords: *ANCA associated vasculitis, Cryoglobulinemia, small vessels vasculitis*

[Abstract:2428]

DERMATOMYOSITIS: SOMETIMES IMMUNOSUPPRESSION MAY NOT BE SUFFICIENT

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Case Description: We report the case of a 73-year-old woman admitted from the emergency department for a three-week history of fatigue, weakness, and rash. Physical examination revealed proximal muscle weakness, heliotrope erythema, rash on the neckline, and lesions consistent with Gottron's papules. Urgent laboratory results showed creatine kinase (CK) levels of 19,759 U/L (normal range: 26-192 U/L), LDH of 524 U/L (normal range: 120-246 U/L), and elevated transaminases.

Diagnostic Pathways: Upon suspicion of dermatomyositis (DM), the investigation was completed with electromyography (normal), autoimmune markers (positive anti-TIF1γ), and skin biopsy (consistent with DM). Treatment was initiated with prednisone and azathioprine, but the patient experienced a sluggish progression, advancing to tetraparesis with severe dysphagia, necessitating gastrostomy. Immunoglobulin cycles were initiated, achieving suboptimal disease control.

However, two weeks later, the patient developed pleural effusion, prompting thoracentesis and cellular block analysis. The pathological findings from the cellular block were consistent with high-grade serous ovarian tumor, further confirmed by biopsy of a newly appearing accessible lymph node. Following the initiation of antineoplastic treatment the patient experienced a remarkable improvement, regaining autonomy, the ability to swallow, and the capacity to walk unassisted.

Discussion and Learning Points: Dermatomyositis exhibits a high rate of association with neoplastic diseases, necessitating thorough screening, especially in patients over 60 years of age and those with positive anti-TIF1γ antibodies. Neoplastic conditions may precede or follow inflammatory pathology. Antineoplastic treatment should be the primary approach, although challenges in diagnosis, particularly when its presence is not evident, constrain the choice of the initial treatment.

Keywords: *dermatomyositi, cancer, immunosuppression*

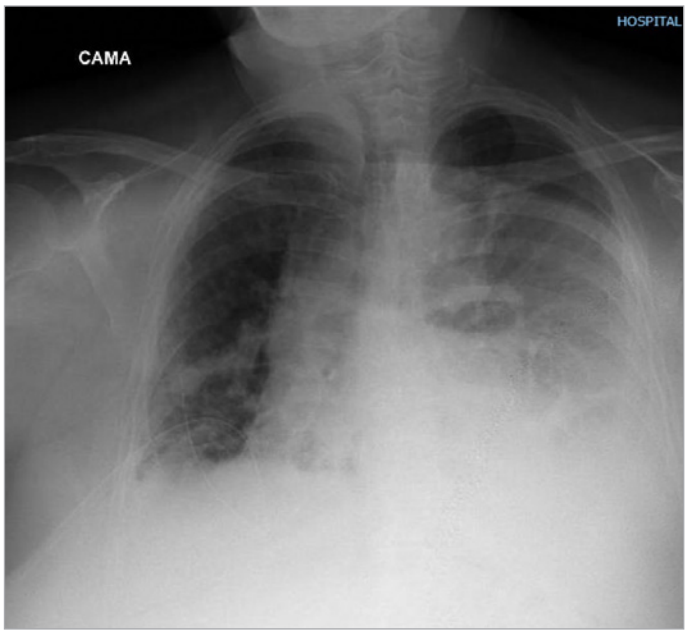


Figure 1. *Pleural effusion.*

[Abstract:2431]

THE EFFECT OF METABOLIC SYNDROME COMPONENTS WITH KNEE OSTEOARTHRITIS

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Purpose: to evaluate the effect of the number of metabolic syndrome (MetS) components on the clinical manifestations of knee osteoarthritis (OA).

Methods: The prospective study included 153 patients with knee OA (ACR) of X-ray stage I-III (Kellgren-Lawrence) and MetS components (NCEP ATPIII), who signed an informed consent. An individual card was filled out for each patient, including clinical and anamnestic data, the results of a clinical examination, an assessment of knee pain according to VAS, and WOMAC indicators. The patients underwent laboratory examination (biochemical blood tests: glucose, total cholesterol, LDL, HDL, TG).

Results: OA patients with the accumulation of two or more MetS components had a longer OA duration, higher intensity of knee pain according to VAS, worse indicators of total WOMAC and its components (Table 1). Patients with two or more MetS components were significantly more likely to take NSAIDs ($p<0.05$). Spearman

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correlation analysis showed significant relationships between the number of MetS components and the duration of OA (r=0.56, p<0.05), age (r=0.57, p<0.05), BMI (r=0.48, p<0.05), knee pain intensity according to VAS (r=0.36, p<0.05), WOMAC indicators (r=0.46, p<0.05).

Conclusions: In our work, it was demonstrated that the accumulation of two or more MetS components is associated with more severe clinical manifestations of knee OA.

Keywords: *osteoarthritis, metabolic syndrome, pain*

Parameters	A group (1 MetS component, n=53)	B group (2 MetS components, n=46)	C group (3 MetS components, n=44)	D group (4 MetS components, n=10)	p (between groups A and B,C, D)
Age, y., Me	54 [46;57]	57.5 [46;65]	60 [46;65]	65 [46;65]	<0.05
Duration of OA, y., Me	4 [8;5.9]	8 [2;15]	15.5 [2;15]	16 [2.1;20]	<0.05
VAS pain, mm, Me	22 [10;45]	30 [11.5;40]	40 [15;60]	45 [15;60]	<0.05
WOMAC pain, mm, Me	139 [50;180]	159 [60;200]	180 [80;220]	215 [80;220]	<0.05
WOMAC functional insufficiency, mm, Me	395 [120;780]	593 [250;790]	740 [300;810]	675 [462;970]	<0.05
Total WOMAC, mm, Me	560 [220;1070]	778 [330;1090]	970 [460;1120]	965 [680;1360]	<0.05

Table 1. *The effect of the number metabolic syndrome components with knee osteoarthritis.*

[Abstract:2444]

WHEN A RASH TAKES YOUR BREATH AWAY: A CASE OF ANTI-MDA5 DERMATOMYOSITIS

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Introduction: Anti-MDA5 dermatomyositis is a rare autoimmune disease that manifests clinically with amyopathic dermatomyositis and rapidly progressive interstitial lung disease.

Case: A 24-year-old male presented with a purpuric rash of the extremities of two weeks' duration. This was preceded by fever, flu-like illness, and dark urine. Prior to that he had a month-long history of malaise, weight loss, and sweating. He was admitted for workup of possible vasculitis. During hospitalization, he was evaluated by a multi-disciplinary team. CT scan of the chest was negative for pulmonary embolism but revealed numerous patchy ground-glass opacities. CT abdomen revealed multiple areas of mediastinal, axillary, and abdominal lymphadenopathy.

Lab results were positive for ENA, SSA, Ro 52 antibody, and MDA5 consistent with a diagnosis of dermatomyositis. He was started on steroid course. For definitive management, the patient received a 1-gram dose of cyclophosphamide and tacrolimus. Unfortunately, despite all of these efforts, his respiratory status continued to deteriorate and he required intubation. The patient was then transferred to another facility for extracorporeal membrane oxygenation (ECMO) support.

Discussion and Conclusions: Anti-MDA 5 is a rare systemic autoimmune disease which carries a guarded prognosis especially with the presence of interstitial lung disease. Despite aggressive upfront therapeutic interventions, there is a high likelihood of treatment failure. A combination of steroids, tacrolimus and cyclophosphamide showed significant improvements in 6-month survival rates (89% vs 33%). (2)

Early recognition and the use of aggressive immunosuppressive drugs are the key factors for improving the survival in patients with Anti-MDA 5 dermatomyositis.

Keywords: *MDA5 dermatomyositis, interstitial lung disease, ECMO*

[Abstract:2480]

ASEPTIC MENINGITIS AS A FIRST PRESENTATION OF CONNECTIVE TISSUE DISEASE

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A 37-year-old female patient, with a recent diagnosis of fibromyalgia, presented with fever and headache, starting twelve days earlier. The patient was alert and febrile. The physical examination revealed neck stiffness. Head Computed Tomography (CT) scan showed no abnormalities. Cerebrospinal Fluid (CSF) analysis revealed 678 white cells, 100% lymphocytes, Hypoglycorrhachia (CSF glucose 29 mg/dl) and protein 280 mg/dl. Dexamethasone, vancomycin, meropenem and acyclovir were started for possible bacterial or viral meningitis and antibiotics were discontinued once the blood and CSF cultures, CSF Gram stain and Polymerase chain reaction tests were negative. One day later, fever recurred and repeat CSF analysis revealed 410 white cells, 100% lymphocytes, Hypoglycorrhachia (CSF glucose 56 mg/dl) and protein 136 mg/dl. Antibiotics and dexamethasone were restarted with immediate clinical response.

The differential diagnosis was bacterial, viral or aseptic meningitis of noninfectious etiology.

The laboratory analysis of blood and CSF ruled out an infectious etiology. Brain CT scan ruled out venous sinus thrombosis and chest and abdomen CT scans did not reveal cancer. CSF immunophenotyping was negative for clonality. The immunological tests revealed Antinuclear Antibodies 1:640 and low positive Anticardiolipin IgM. She was treated with pulse steroid intravenous therapy by rheumatologists and the patient fully recovered. The most possible diagnosis was considered to be aseptic meningitis as an initial clinical presentation of connective tissue disease. In the follow up, tapering doses of oral steroids were continued with the addition of azathioprine.

Aseptic meningitis related to autoinflammatory conditions should be considered after careful exclusion of other etiologies.

Keywords: *aseptic meningitis, fever, connective tissue disease*

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[Abstract:2485]

AN UNUSUAL MODE OF REVELATION OF AN EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA) CO-EXISTING WITH A SJOGREN'S SYNDROME (SS)

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Anti-Neutrophil Cytoplasmic Antibodies (ANCA)-associated vasculitides (AAV) encompass a group of rare auto-immune diseases characterized by granuloma formation and/or inflammation of small vessels. The clinical spectrum of AAV varies widely. The liver location is exceptional leading to diagnostic challenges. Moreover, AAV can co-occur with other systemic diseases, further complicating the diagnosis. We herein present a unique case of AAV overlapping with Sjogren's syndrome (SS), with an uncommon mode of revelation.

A 47-year-old female, was admitted for hiatal hernia surgery. During the intervention, nodular hepatomegaly was discovered. A liver biopsy was performed showing non-necrotizing epithelioid and central giant cell granulomas. Computed-Tomography (CT) scan showed perilymphatic pulmonary micronodules with bilateral hilar lymphadenopathies, raising the suspicion of sarcoidosis. Minor salivary gland biopsy (MSGB) revealed a Chisholm grade 3 sialadeinitis, that along with the patient's dry eye and mouth symptoms confirmed Sjogren's syndrome. Immunological workup showed negative Antinuclear antibodies (ANA) and positive anti-myeloperoxidase (MPO) antibodies. A year later, the patient presented with asthma flare-ups and ENT symptoms. A nasal biopsy was performed showing signs of Eosinophilic Leukocytoclastic Vasculitis, confirming the diagnosis of Eosinophilic Granulomatosis with Polyangiitis (EGPA). Oral steroid therapy was initiated, resulting in clinical improvement.

In conclusion, we present a case highlighting that EGPA is a protean disease, possibly mimicking or co-occurring with other auto-immune disorders. Thus, the importance of considering the differential diagnoses and carefully monitoring for any new symptom or organ dysfunction.

Keywords: *eosinophilic granulomatosis with polyangiitis, hepatic granuloma, ANCA associated vasculitis, Sjogren's syndrome, late onset asthma*

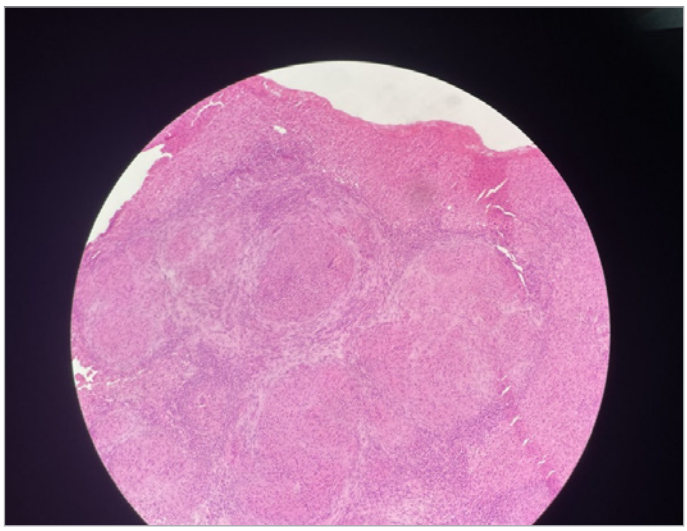


Figure 1. *Liver Biopsy.*
Non-necrotizing epithelioid and central giant cell granulomas of the liver.

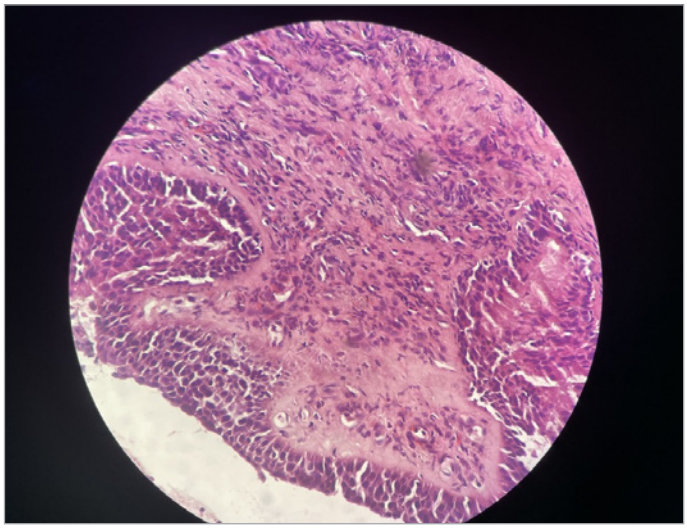


Figure 2. *Nasal biopsy.*
Eosinophilic Leukocytoclastic Vasculitis.

[Abstract:2513]

DIFFUSE INFILTRATIVE LUNG DISEASE IN AN INTERNAL MEDICINE DEPARTMENT, ABOUT 160 CASES

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Background: The etiologies of diffuse infiltrative lung disease (DILD) are diverse. The etiological investigation is guided by the radiological pattern, extra-pulmonary manifestations and immunological and anatomopathological investigations.

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Patients and Methods: This is a descriptive, retrospective study carried out between 2015 and 2023, including 160 cases of DILD. The diagnosis of PID was based on high-resolution thoracic CT scan.

Results: There were 115 women and 45 men. The average age was 58. A radiological pattern was identified in 116 patients. These were UIP (n=33), PINS (n=17), fibrosing PINS (n=13), organized pneumonia (n=11, sarcoidosis (n=41) and silicosis pattern (n=1). Extra-respiratory manifestations were present in 108 patients (67.5%). Periungual capillaroscopy showed major scleroderma dystrophies in 13 patients. Labial biopsy showed Chisolm stage ≥3 lymphocytic sialadenitis (n=29), granuloma without caseous necrosis (n=2) and localized amyloidosis (n=2). Bronchial biopsies showed granuloma without caseous necrosis in 18 cases (31.5%). Anti-nuclear antibodies were positive in 40% of patients. The etiology of DILD was identified in 113 patients (70%). The etiology was a connective tissue disease (n=47), including 14 cases of primary Sjogren's syndrome, 14 cases of inflammatory myopathy, 8 cases of rheumatoid arthritis, 2 cases of systemic lupus erythematosus, and 2 cases of scleromyositis. Other etiologies were sarcoidosis (n=40), IPAF (interstitial lung disease with autoimmune manifestations) (n=9), ANCA vasculitis (n=6), IRS sequelae of covid-19 lung disease (n=8), other causes (n=4).

Conclusions: This study is characterized by its etiological diversity, with a predominance of sarcoidosis and connective tissue diseases.

Keywords: *diffuse infiltrative lung disease, sclerodermia, Sjogren syndrome*

[Abstract:2517]

AN UNUSUAL DIAGNOSIS AT THE BEDSIDE

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Clinical Case: This is an 85-year-old man with atrial fibrillation who was anticoagulated with apixaban that came to the emergency department due to pain and functional impotence in his right hand. On examination, there was swelling of the hand and small palpable reddish lesions on both legs that did not disappear with pressure, compatible with petechiae.

Diagnostic Pathways: Blood and urine tests were performed, which revealed a slight deterioration in kidney function with creatininemia of 1.3 mg/dL (upper limit of normal in 1.2 mg/dL), C-reactive protein levels of 100 mg/l (upper limit of normal in 5 mg/L) with normal procalcitonin levels, D-dimer in 2.87 mg/dL (upper limit of normal in 0.5 mg/dL) and microscopic hematuria. We performed a bedside ultrasound examination of the right upper limb, ruling out venous thrombosis, but seeing signs compatible with tenosynovitis of the flexor muscles of the right hand, in the form of swelling of the sheath (left image, yellow arrows) with increased signal color Doppler (right image). Finally, hospital admission was decided and a study was completed with a skin biopsy, in which immunoglobulin A deposits were observed in superficial vessels. With all these data, the diagnosis of Henoch Scholein purpura was assumed and boluses of methylprednisolone were prescribed, with subsequent transition to oral prednisone and mycophenolate mofetil. After resolution of the tenosynovitis, deterioration of renal function and hematuria, the patient was discharged.

Conclusions: In this case, the bedside ultrasound performed by the internal medicine team allowed the diagnostic process to be accelerated.

Keywords: *ultrasound, kidney, skin*

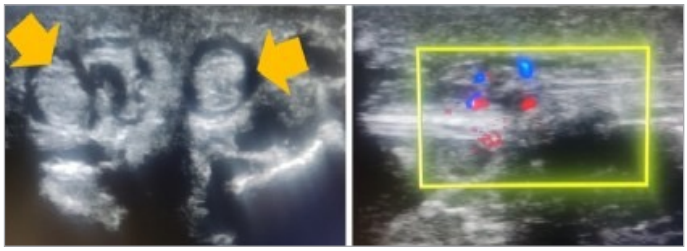


Figure 1.

[Abstract:2519]

A CASE OF BEHÇET'S DISEASE PRESENTING WITH PORTAL VEIN AND HEPATIC VEIN THROMBOSIS

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Case Description: A 20-year-old male patient with known diagnoses of Behçet's disease and epilepsy was admitted to the emergency department with abdominal pain and distension that has been ongoing for a month. His laboratory test revealed total bilirubin of 2.79 mg/dl, direct bilirubin of 2.24 mg/dl, and INR of 1.7. Portal doppler ultrasonography was performed after 9 cm of fluid was seen in the abdomen during abdominal ultrasonography. Hepatic and portal vein thrombosis was detected. Follow-up was started for initial diagnosis of Budd-Chiari syndrome secondary to Behçet's disease.

Clinical Hypothesis: Multiple thrombi in a young patient diagnosed with Behcet's disease, Budd-Chiari syndrome.

Diagnostic Pathways: The patient had coexisting hepatic and portal vein thrombosis. Abdominal CT angiography was performed and thrombus was confirmed. He was diagnosed with Budd-Chiari syndrome secondary to Behçet's disease. The patient developed confusion during the process, so cranial imaging was performed for neuroBehçet's disease, and it was found to be normal. The patient was simultaneously evaluated for hepatic encephalopathy.

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He developed flapping tremor and his ammonia level was detected as 163 µmol/L, thereby hepatic encephalopathy treatment started immediately. Unfortunately, the patient died due to liver failure.

Discussion and Learning Points: Behçet's disease is an auto-inflammatory systemic vasculitis that is most commonly diagnosed between the ages of 20-30, has a similar incidence in men and women, but is more mortal in men. It can affect all vessels although multiple thrombi simultaneously are less common, they can involve the hepatic and portal veins and cause Budd-Chiari syndrome.

Keywords: *Behçet's disease, Budd-Chiari, hepatic encephalopathy*

[Abstract:2527]

EOSINOPHILIC FASCIITIS

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Patient aged 56 years old referred to the hospital to study persistent eosinophilia and myalgias, diffuse swelling/skin hardening in the forearms. No Raynaud. At examination he showed no arthritis, tendon retraction in flexors of 3rd/5th fingers in arms, cutaneous sclerosis in arms/lower limbs, orange peel. Tests performed were: eosinophilia, negative autoimmunity; MRI: signs of subfascial muscle edema in lateral vastus, semitendinosus and extensor digits (findings consistent with eosinophilic fasciitis). Muscle biopsy: inflammatory infiltrates at septal level, atrophy and muscular necrosis. ENG: normal. Capillaroscopy: normal. Hematologic process was ruled out and the diagnosis was eosinophilic fasciitis. He was initially treated with prednisone, later methotrexate with good response. Eosinophilic fasciitis is an uncommon scleroderma-like syndrome. The cutaneous involvement is similar to scleroderma spectrum syndromes; however, sclerodactyly is absent. The groove sign is characteristic: a visible indentation in the course of the veins. Most patients have transient blood eosinophilia, not correlated with severity. EF is associated with check-point inhibitors, autoimmune diseases or a form of presentation of hematologic processes. The diagnosis is confirmed with skin-to-muscle biopsy (no pathognomonic signs). Magnetic resonance imaging is useful for diagnosis and to guide biopsy. Initial management is with glucocorticoids at doses equivalent to 1 mg/kg/day and doses are reduced when skin improves, this may take months. Underlying cause should be treated. Subsequent therapy: methotrexate (15-25 mg/week) and alternatives are mycophenolate or hydroxychloroquine. 75% respond to glucocorticoid therapy and 50% achieve remission. Cutaneous truncal involvement is associated with resistance to treatment. JAK inhibitors, anti TFN open a window of hope for refractory disease.

Keywords: *eosinophilic fasciitis, scleroderma-like syndrome, methotrexate*

[Abstract:2546]

SYNCOPE AS A MANIFESTATION OF ERYTHEMATOSUS SYSTEMIC LUPUS FLARE IN A YOUNG WOMAN

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Summary: Patient with a history of systemic lupus erythematosus (SLE) who consulted for syncope in the emergency room. Complementary tests are performed with findings of hemolytic anemia and ischemic stroke.

Purpose: To present the case of a patient with a history of ESL and neurological symptoms.

Case Report: We report the case of a 35-year-old woman with a personal history of erythematosus systemic lupus and obstetric antiphospholipid syndrome without treatment due to abandonment. The patient consulted at the emergency room referring loss of consciousness and fall on that day accompanied by asthenia in the previous 4 days. No fever or other symptoms.

Findings: At physical examination blood pressure was 130/70 mmHg, heart rate 80 bpm, no fever. Cardiopulmonary auscultation was normal and there was no neurological focality. Lymphadenopathy was palpable in the left axilla.

Blood test: Hb 6 g/dl, leukocytes 10300/L, platelets 309000/L, direct coombs test was positive, reticulocytes 11.5%, peripheral blood smear: anisocytosis and polychromasia. Creatinine 1 mg/dl, RCP 11 mg/L, ESR 92 mm/h. C3 67, C4 <2.9. ANA, lupic anticoagulant and Anti-B2M antibody immunoglobulin G were positive. Coagulation, liver enzymes and ionogram were normal.

Cranial TC-scan: small right paraventricular hypodense focus in relation to ischemic stroke (which is confirmed by MRI). Thorax RX: no findings. Electrocardiogram: sinusal rhythm without repolarization disorders. The patient received treatment with intravenous methylprednisolone boluses, oral hydroxychloroquine, and subsequently rituximab. During admission, positive serology for hepatitis B virus was received, so treatment with tenofovir was started.

Conclusions: Early detection of flares in patients with ESL is important for early management and avoid complications.

Keywords: *ESL, antiphospholipid syndrome, anemia, syncope*

[Abstract:2547]

SAPHO SYNDROME REVEALED BY BILATERAL UVEITIS

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Introduction: SAPHO syndrome is a rare entity, of recent individualization which combines: synovitis, acne, palmoplantar

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pustulosis, hyperostosis, osteitis. Ocular manifestations are rarely revealing. The enteropathogenic mechanism remains poorly elucidated. Given its clinical-biological and radiological heterogeneity, difficult diagnosis, it is based on a set of criteria. We report the case of a patient hospitalized in internal medicine.

Observation: We report the case of a 54-year-old patient, with acute bilateral anterior uveitis revealed by eye pain and reduced visual acuity, associated with bilateral ankle arthritis. The patient had a history of palmoplantar pustulosis. The symptomatology had been evolving for a month marked by peripheral and axial polyarthritis, painful swelling of the sternoclavicular joint. Patient was afebrile and in conserved general condition. The biological and inflammatory assessment were positive however immunological was normal, the search for HLA B27 and B51 were negative. Erosive and condensing arthritis of the manubriosternal bone were revealed by the CT scan, with scintigraphy hyperfixation on the side, allowing the diagnosis of SAPHO to be made. Corticosteroid therapy was effective after 6 months.

Conclusions: SAPHO syndrome is an underdiagnosed systemic condition, integrated into spondyloarthropathy. The reported ocular damage is rare but serious, compromising the visual prognosis. The recognition of this syndrome as a pathological entity in its own right has enabled considerable therapeutic progress.

Keywords: *SAPHO, uveitis, spondyloarthropathy*

[Abstract:2560]

MICROSCOPIC POLYANGIITIS

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A 40-year patient came to emergency due to progressive dyspnea and polyarthralgias. At examination the patient showed no arthritis, bibasal crepitant, palpable purpura in lower limbs. At tests the patient reported elevation of acute phase reactants, positive pANCA autoimmunity. Chest CT revealed pulmonary hemorrhage. Skin biopsy showed leukocytoclastic vasculitis. ENG was normal. The infectious process was ruled out and diagnosis was MPA. The patient was treated with corticosteroid pulses and later cyclophosphamide NIH scheme with good response. Maintenance treatmen was with rituximab.

Antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides (AAV) include granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), limited renal vasculitis, and eosinophilic granulomatosis with polyangiitis (EGPA). They affect small arteries and have similar histology in the kidney.

Patients present nonspecific symptoms: fever, anorexia, weight loss, arthromyalgia. Organ-specific manifestations include: Otolaryngologic manifestations: nasal crusting, sinusitis, otitis media, persistent rhinorrhea, purulent/bloody nasal discharge, ulcers, polychondritis, hearing loss. Lung involvement: hoarseness, cough, dyspnea, hemoptysis or pleuritic pain. A chest radiograph may reveal pulmonary nodules, opacities and infiltrates. Typical renal presentation are rapidly progressive glomerulonephritis (focal necrotizing pauciimmune). Cutaneous manifestations are purpura in lower extremities with/without focal necrosis/ulceration. Ophthalmic involvement include conjunctivitis, episcleritis/scleritis, optic neuropathy, retinal vasculitis and uveitis, retroorbital pseudotumor. Neurological manifestations are mononeuritis multiplex, cranial nerve abnormalities. Diagnosis is based on clinical findings (constitutional symptoms and possible glomerulonephritis or upper/lower respiratory tract involvement), laboratory tests (positive ANCA but false positives/negatives exist), imaging studies and histological examination (kidney/skin/lung) are the most definitive method.

Differential diagnosis is a challenge. Mimickers are EGPA, polyarteritis nodosa, anti-glomerular basement membrane antibody (anti-GBM) disease, drug/cocaine-induced ANCA-associated vasculitis, infection, and malignancy.

Keywords: *microscopic polyangiitis, pulmonary hemorrhage, pANCA*



Figure 1. *After treatment.*

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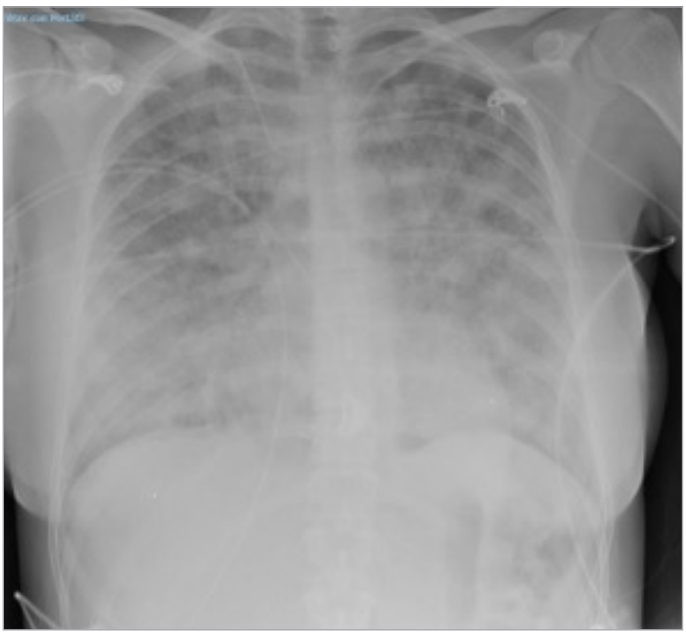


Figure 2. Before treatment.

[Abstract:2582]

USE OF CAROTID ULTRASOUND TO ASSESS CARDIOVASCULAR RISK IN PATIENTS DIAGNOSED WITH SYSTEMIC SCLEROSIS AND SYSTEMIC LUPUS ERITHEMATOSUS

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Purpose: The main purpose of this study is to evaluate whether carotid ultrasound (US) is a useful tool in global cardiovascular (CV) risk assessment in patients with Systemic Sclerosis (SSc) and Systemic Lupus Erythematosus (SLE).

Methods: We selected 31 patients under 70 years, with SSc or SLE. All of them underwent a basic evaluation with anthropometry, blood pressure and a structured clinical interview. SCORE-2 was used to calculate the CV risk. In addition, carotid US was performed in all patients by trained physicians to evaluate the presence of significant atheroma plaques (>1.5 mm).

Findings: The study included 31 patients, 15 of them diagnosed with SSc and 16 with SLE. In a basic evaluation using SCORE-2, 13.3% and 30.8% were classified as low CV risk and 86.7 and 69.2% as moderate CV risk, respectively for the SSc and SLE groups. We found significant plaques in 33.3% of patients with SS and 28.5% with SLE. Considering the US results, these patients could be re-stratified to very high CV risk according to the European Society of Cardiology guidelines.

Conclusions: In almost one third of our patients significant plaques were found. The finding of carotid plaques with US was established by the European Society of Cardiology guidelines of 2021 as a CV risk modifier, restratifying patients with carotid atheroma plaques as very high CV risk.

Reference:

L.J. Visseren F. et al. 2021 ESC Guidelines on cardiovascular disease prevention in clinical practice. 2022 75(5):429.e1-429.e104.

Keywords: *systemic sclerosis, systemic lupus erithematosus, cardiovascular risk, carotid ultrasound.*

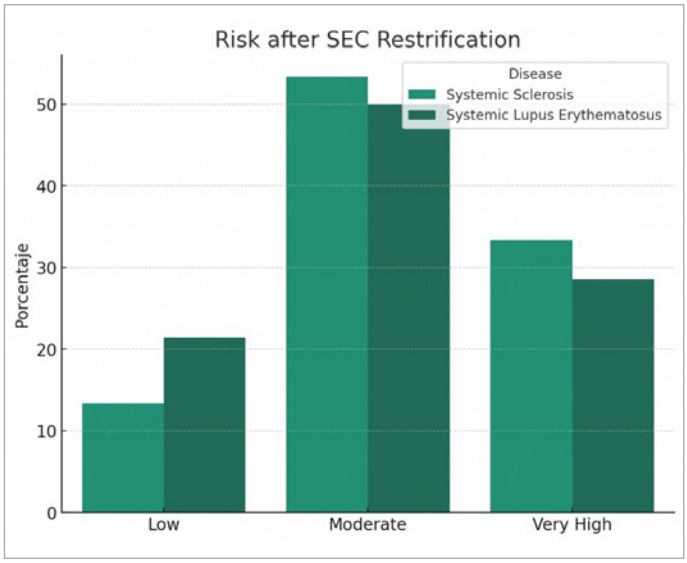


Figure 1. Cardiovascular Risk of patients after restratification according to ESC 2021 criteria.

[Abstract:2598]

CLINICAL RELEVANCE OF ANTI-CENTROMERE ANTIBODIES POSITIVITY IN ROUTINE PRACTICE

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Anti-centromere antibodies (ACA) belong to antinuclear antibodies (ANA) and are reported to be associated with systemic sclerosis. ACA can be detected by indirect immunofluorescence (IIF) (particular staining pattern) or by immunodot (ID) using CENP-B antigen.

We aimed to evaluate IIF/ID concordance, ACA positivity frequency and its clinical relevance in routine practice.

We conducted a retrospective study (18-months) concerning patients who underwent ANA testing by IIF on Hep-2 cells and ID (Euroimmun®). Those who had positive ACA by IIF and/or ID were included. Clinical data of patients from internal medicine department were collected.

ACA were detected in 39 cases (1,8 % of ANA testing requests (IIF+ID) during the study period): mean-age=40.5years (3-65); sex-ratio=3men/36women; requests from internal medicine department=35.8%. ANA-IIF titers ranged from 1/160 (2.5%) to

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≥1/1280 (61.5%). Concerning ACA detection, IIF and ID results were concordant in 12 cases (30.7%). Discordant results included 2 IIF+/ID- cases and 25 IIF-/ID+ cases (IIF-speckled pattern / euroline signal: 17 cases (+), 2 cases (++), 6 cases (+++)). No ANA antigenic specificity (other than CENP-B) was detected by ID in 21 cases. In the remaining cases, anti-SSA (n=5), anti-Ro52 (n=5) and anti-Sm/RNP (n=4) were the most frequent associated specificities.
Among the 14 patients with available clinical data, no systemic sclerosis case was identified. Other connective tissue diseases (systemic lupus erythematosus, dermatomyositis, mixed connective tissue disease) were diagnosed in only 4 cases.
IIF represents a first-line, operator-dependent technique for ANA detection while ID is useful to confirm antigenic specificity. ACA are not specific of systemic sclerosis. Interpretation of these tests' results relays mainly on clinical context.

Keywords: *anti-centromere antibodies, systemic sclerosis, indirect immunofluorescence, immunodot*

[Abstract:2606]

NEUROLOGICAL MANIFESTATIONS REVEALING SJORGEN'S SYNDROME, ABOUT 5 CASES

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Introduction: The neurological involvement in Sjogren's syndrome(SS) is polymorphic with disastrous outcomes Their frequency is variously appreciated. Peripheral involvement is by far the most studied with a frequency of 10 to 30%. Central involvement is rarer and affects less than 5% of patients. We report a series of 5 cases of patients presenting neurological symptoms revealing SS.
Methods: This is an observation made from medical reports of patients hospitalized in our service for the management of neurological disorders revealing SS.
Results: All our patients were female with an average age of 46.5 years. The diagnosis time was 31 months. SS was primary in 60% of cases. The neurological manifestations were revealing in all cases. Peripheral neuropathy was more frequent and polymorphic with sensory-motor polyneuropathy, proprioceptive ataxia, vestibular syndrome and bilateral sensorineural deafness. The central involvement was an intracranial hypertension syndrome in one patient, a cerebellar syndrome in another. Brain MRI showed white matter lesions in both cases. The antinuclear antibody test (ANA) was positive in three patients. A biopsy of accessory salivary glands was contributory in 80% of cases. All patients received treatment with corticosteroid therapy. The use of immunosuppressive treatment was necessary.
Conclusions: Neurological involvement can reveal SS in 10 to 15%. Consider this syndrome in front of any unexplained neurological manifestations. The difficulty in diagnosing these disorders is the cause of a diagnostic delay.

Keywords: *Gougerot-Sjogren, neurology, prognosis, inaugural*

[Abstract:2613]

RENAL THROMBOSIS AND ANTIPHOSPHOLIPID SYNDROME. CHANCE OR CASUALITY?

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We report the case of a 73-year-old woman with good quality of life, hypertension (HT), diabetes mellitus type 2 (T2TM), chronic renal disease (CKD) stage IIIa, and follow-up by cardiology for alternating episodes of atrial fibrillation (AF) and atrial flutter with no evidence of structural heart disease. She presented herself to the ER for fever and subacute abdominal pain, no apparent infectious focus or epidemiological history of interest. She was diagnosed with bilateral renal infarction by means of computed axial tomography (CT) after two months of underanticoagulation due to a traumatic acute subdural hematoma. The extension of the thrombotic event made us suspect the possibility of another underlying cause, expanding the study of hypercoagulability that evidenced the presence of a triple positive primary antiphospholipid syndrome not filiated so far.
Antiphospholipid syndrome is a systemic autoimmune disease with high thromboembolic potential and high morbidity and mortality, especially in those patients not diagnosed and therefore not anticoagulated. The heterogeneity of the clinical manifestations, the laboratory data and the associated intrinsic or extrinsic factors make it an entity of great impact at the multisystemic level. Therefore, evaluating the prognosis in these patients through disease activity, organic damage and quality of life is crucial in clinical practice. The diagnosis remains a medical challenge but clinical manifestations have been described "extra criterion", which although not part of the classification criteria are of vital relevance when taking a therapeutic behavior in front of patients in which it is suspected.

Keywords: *antiphospholipid syndrome, kidney infarction, hypercoagulability*

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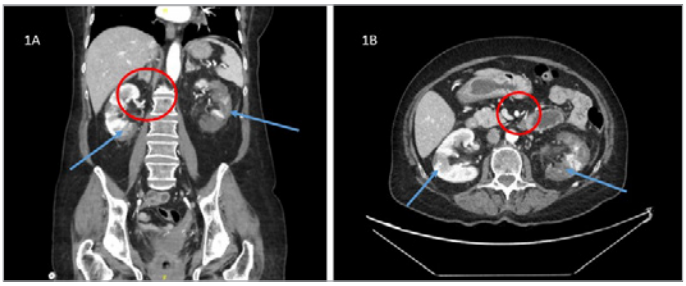


Figure 1. Computed axial tomography of kidney lesions.
1A) Axial cut of abdominal computed tomography. 1B) Comparative coronal cut. In both figures the arrows show corticomedular hypodensity compatible with renal ischemia. The circle locates the thrombus at the level of the superior mesenteric artery.

Analytics in emergencies	Analytics in hospitalization	Analytics at discharge	Analytics in consultation
Hemogobin:10.3 g/dl	Hemogobin:9.6 g/dl	Hemogobin:10 g/dl	Hemogobin:10.1 g/dl
Leukocytes:22.410 x10^9/L	Leukocytes:17.003 x10^9/L	Leukocytes:15.150 x10^9/L	Leukocytes:8740 x10^9/L
Neutrophils:19.340 x10^9/L	Neutrophils:13.005 x10^9/L	Neutrophils:12.000 x10^9/L	Neutrophils:5.770 x10^9/L
Monocytes:1.790 x10^9/L	Monocytes:1.113x10^9/L	Monocytes:0.900x10^9/L	Monocytes:0.500 x10^9/L
Platelets:329.000x10^9/L	Platelets:523.000x10^9/L	Platelets:600.000x10^9/L	Platelets:512.000x10^9/L
Prothrombin activity: 67%	Prothrombin activity: 93%	Prothrombin activity: 100%	Prothrombin activity: 37%
INR: 1.29	INR: 1.05	INR: 1.	INR: 2.19
Fibrinogen: 986 mg/ dL	Fibrinogen: 1125 mg/ dL	Fibrinogen: 1138 mg/ dL	Fibrinogen: 571 mg/ dL
Glucose:238 mg/dl	Glucose:78 mg/dl	Glucose:271 mg/dl	Glucose:114 mg/dl
Urea:36 mg/dl	Urea:61 mg/dl	Urea:25 mg/dl	Urea:32 mg/dl
Creatinine:1.27 mg/dl	Creatinine:1.60 mg/dl	Creatinine:1.16 mg/dl	Creatinine:1.10 mg/dl
GFR:42 ml/min/1.73m3	GFR:32 ml/min/1.73m3	GFR:47 ml/min/1.73m3	GFR:48 ml/min/1.73m3
LDH:2090 UI/L	LDH:965 UI/L	LDH:616 UI/L	LDH:148 UI/L
GOT:77 UI/L	GOT:29 UI/L	GOT:14 UI/L	GOT:9 UI/L
GPT:145 UI/L	GPT:39UI/L	GPT:23 UI/L	GPT:7 UI/L
CRP:353.5 mg/L	CRP:315 mg/L	CRP:108 mg/L	CRP:1.9 mg/L

Table 1. Comparative results of the analytics.
INR: International Normalized Ratio; GFR: glomerular filtration rate; LDH: lactate dehydrogenase; GOT: glutamic oxalacetic transaminase; GPT: glutamic pyruvic transaminase, CRP: C-reactive protein

[Abstract:2645]

A RARE PRESENTATION OF ADULT-ONSET STILL'S DISEASE IN A 74-YEAR-OLD FEMALE

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Introduction: Adult-Onset Still's Disease (AOSD) is a systemic inflammatory disorder. The diagnosis is one of clinical suspicion, requiring the exclusion of infection, malignancy, and systemic disease. This case report details the clinical presentation, diagnostic challenges, and management of a 74-year-old female with AOSD.

Observation: The patient presented with persistent fever, polyarthralgia, and salmon pink back rash, initially posing a diagnostic dilemma. Laboratory investigations revealed raised leucocytosis with neutrophil predominance, marked hyperferritinemia associated with diminished concentration of glycosylated ferritin, and elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). The rheumatoid factor (RF), anti-nuclear antibody (ANA), and anti-dsDNA, blood and urine cultures, and tumor markers were also negative, thoraco abdomino pelvic CT scan was normal, treatment of this patient consisted of corticosteroids, disease modifying anti-rheumatic drugs (DMARD) -methotrexate-, the patient is currently in remission.

Conclusions: The report highlights the atypical onset of Still's disease in the elderly, emphasizing the importance of considering AOSD in the differential diagnosis of fever of unknown origin in older individuals. Timely recognition and appropriate management are crucial for improving outcomes in this rare manifestation of Still's disease.

Keywords: *adult-onset Still's disease, elderly, fever*

[Abstract:2656]

KEEP IN MIND! DRUG-ASSOCIATED VASCULITIS: ZOLEDRONIC ACID-INDUCED LARGE VESSEL VASCULITIS DIAGNOSED BY COMPUTERIZED TOMOGRAPHY

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Introduction: Certain medications may induce forms of vasculitis. Here we report a case diagnosed with Large Vessel Vasculitis(LVV) induced with zoledronic acid (ZolAc) infusion.

Case: A 50-year-old postmenopausal woman diagnosed with hypertension and osteoporosis (OP) was admittted to our department complaining of a 4-week history of fever up to 40°C, flu-like symptoms, myalgia and arthralgia. She was treated for OP with intravenous infusion of ZolAc 5 mg for the first time, four weeks before.The day after infusion, she started to have a persistent fever of 39°C and above, which never decreased. Due to swelling and pain in her left ankle, the blood tests taken revealed elevated acute phase reactants (AFR); Erythrocyte Sedimentation Rate (ESR)-100 (0-25 mm/h) and C-reactive Protein (CRP)-145 (<5 mg/L). Patient was prescribed cephalosporin and ibuprofen. She was admitted to our department because her symptoms did not fully improve but tending to decrease in the last 10 days. Body temperature was 37.3°C. Physical examination was completely normal with no organomegalia, arthritis or cutaneous manifestations. AFR were still high but tended to decrease; ESR-53, CRP-24,5 and fibrinogen-583.83(180-350mg/dL). RF, ANA, ANA-IF, ANCA, anti-CCP were negative. Suspicious wall thickening and contrast enhancement compatible with LVV were observed in the ascending aorta, aortic arch, brachiocephalic trunk and proximal to the origin of the left subclavian artery and the descending thoracic aorta on Computerized Tomography (CT) (Figure-1). We decided to follow-up the patient without treatment due to the decrease in complaints and AFR. All symptoms were regressed and AFR decreased to normal levels after 2 weeks

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(ESR;12, CRP;<3.08, fibrinogen 317).

Conclusions: Postinfusion fever, constitutional symptoms, AFR elevation and vasculitis with ZolAc have been reported as a rare complication. LVV should keep in mind as a complication of ZolAc.

Keywords: *zoledronic acid, fever, acute phase reactans, vasculitis, large vessel vasculitis, drug-associated vasculitis*

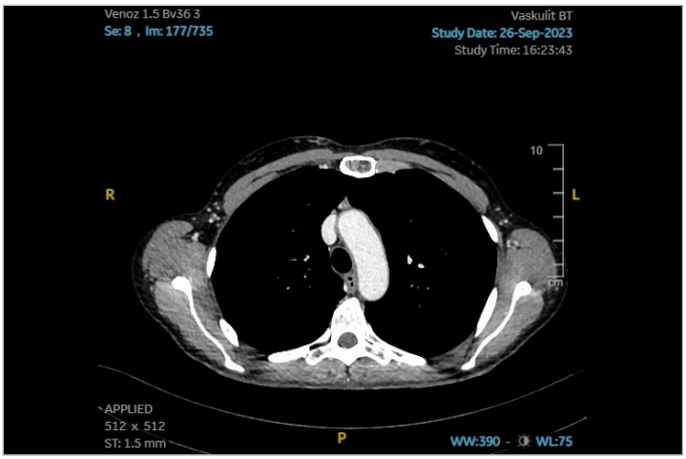


Figure 1. *Wall thickening and contrast enhancement compatible with LVV in the ascending aorta and aortic arch on Computerized Tomography (CT).*

[Abstract:2696]

STUDY OF BONE LESIONS. THE IMPORTANCE OF THE CLINICAL HISTORY

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A 58-year-old female patient with no relevant medical history came for consultation after undergoing an MRI of the shoulder showing patchy bone lesions in the humerus, scapula and clavicles suggestive of metastatic lesions. The patient had no weight loss or alarm symptoms.

Mammography was requested, which showed no alterations, blood tests and bone scintigraphy for better visualization of the bone lesions under study. The scan showed numerous bone erosions in the axial skeleton, iliac crest and humerus, compatible with metabolic bone disease vs. gammopathy. Analytically, there was a monoclonal Ig M Kappa peak, with calcium, total proteins and light chains in blood and urine without alterations. In view of these findings, an iliac crest puncture was performed where one of the bone lesions was located, where no plasma cell proliferation or other findings suggestive of multiple myeloma were observed. With all the study, the diagnosis of probable metabolic bone disease was assumed and close follow-up of the patient was agreed to see the evolution of the bone lesions.

The study of metabolic bone disease is broad and complex. In metabolic pathology, bone scan can show certain abnormal patterns that reflect increased bone metabolism, but they are non-specific and can be observed in any bone hypermetabolic process, regardless of its etiology, so it is important to always rely on the clinical history and examination.

Keywords: *metabolic bone disease, bone lesions, bone scan*

[Abstract:2697]

DYSFERLINOPATHY WITH MYOSITIS APPEARANCE: A CASE REPORT

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A class of uncommon muscular dystrophies known as dysferlinopathy is brought on by recessive mutations in the DYSF gene. The disease may be associated with asymptomatic elevated serum creatin kinase or progressive muscle weakness. They typically begin in mid-to-late childhood or early adulthood. They very seldom become clinically present in old age.

A 53-year-old woman presented to our rheumatology outpatient clinic with a 10-year-long history of progressive muscle weakness and myalgia. She had difficulty eating and drinking water, so she lost 20 kilos in the last 2 years. She has been wheelchair-bound for three months. The patient was started on corticosteroids at another center with a diagnosis of myositis. In the neurological examination of the patient, there was muscle weakness, mostly on the right side. The serum CK level was 500 U/L. The electromyogram showed active myogenic uptake. PETCT revealed that muscular tissues, particularly the muscles of the upper limbs, exhibit increased absorption of FDG.

The left deltoid muscle biopsy showed results consistent with dysferlinopathy. First, we started with a pulse of 1000 mg of methylprednisolone treatment.

The patient, whose symptoms did not subside after methylprednisolone, was given IVIG treatment (15 mg/kg/day for 5 days). MMF (1500 mg/day) was added to the treatment. With the treatment, CK levels were normalized, and muscle strength started to recover slowly.

Keywords: *dysferlin, dysferlinopathy, autosomal recessive muscular dystrophy, myositis*

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[Abstract:2700]

MICROSCOPIC POLYANGIITIS PRESENTING AS A CASE OF FEVER OF UNKNOWN ORIGIN

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We introduce the case of an 82-year-old woman that consulted because of a three-month evolution chart consisting in vespertine fever associated to a general syndrome. In the basic analysis of urgencies, a normocitic anemia and an acute kidney injury were present.

We performed various tests including a quantiferon, a total-body computed tomography scan, different serologies and an autoimmune blood test. It turned to be positive the anti-neutrophil cytoplasmic antibody (ANCA) against the neutrophil proteins leukocyte proteinase 3 (P-ANCA, PR-3).

A biopsy of the kidney was made, that turned to be compatible with microscopic polyangiitis.

We started treatment with methylprednisolone (250 mg per day during 3 days) followed by an infusion of rituximab. Correction of the kidney function occurred, followed by clinical improvement and disappearance of the fever.

The ANCA-associated vasculitides (AAVs) are a group of small-vessel vasculitis characterized by the development of autoantibodies to the neutrophil proteins leukocyte proteinase 3 (PR3-ANCA) or myeloperoxidase (MPO-ANCA). There are three subgroups: granulomatosis with polyangiitis (GPA), microscopic polyangiitis and eosinophilic GPA (EGPA).

Although any tissue can be involved in AAV, the upper and lower respiratory tract and kidneys are most commonly and severely affected.

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Keywords: *Microscopic polyangiitis, Fever of unknown origin, ANCA-associated vasculitis.*

[Abstract:2701]

THROMBOEMBOLIC DISEASE ASSOCIATED WITH SYSTEMIC VASCULITIS AND CONNECTIVE TISSUE DISEASE

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Background: The aim of our study was to describe epidemiological, clinical, paraclinical, therapeutic and outcomes particularities of venous thromboembolic disease (VTE) in patients with systemic vasculitis (SV) and connective tissue diseases (CTD).

Methods: We conducted a retrospective study throughout 19 years of collecting data of patients with SV or CTD and who presented a VTE.

Results: A total of 177 patients were included. Mean age thromboembolic event diagnosis was 38.4 ±13 years. Gender ratio M/F was 1.64. VTE involved 204 locations. Typical sites (lower limb or pulmonary embolism) were described in 84.3% of cases and unusual sites in 15.7%. Three patients had family history of VTE and 28 had personal history of VTE. VTE was associated with SV in 123 patients (69.4%) and CTD in 54 patients (30.5%). SV associated with VTE were Behçet's disease (n=114), microscopic polyangiitis (n=1), cryoglobulinemia (n=1), and giant cell arteritis (n=1). CTD associated with VTE were antiphospholipid syndrome (n=25), systemic lupus erythematosus (n=23), inflammatory myopathies (n=2), Sjogren's Syndrome (n=1) and rheumatoid arthritis (n=1). The others identified thromboembolic risk factors were hyperhomocysteinemia in 21 cases (35.5%) and constitutional thrombophilia in 3 cases. All patients underwent an anticoagulant treatment with a mean duration of 45.6 months. Patients with Behçet's disease also received corticosteroids in 80 cases associated with immunosuppressive therapy in 22 cases. After an average follow-up time of 46.75 months, 42 patients (23.7%) had at least one recurrence of VTE.

Conclusions: VTE is a common complication of Behçet's disease and antiphospholipid syndrome. In the other SV and CTD, screening for others TE risk factors should be performed.

Keywords: *connective tissue disease, vasculitis, thromboembolic disease*

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[Abstract:2753]

THE MORTALITY OF INTERSTITIAL LUNG DISEASE ASSOCIATED WITH CONNECTIVE TISSUE DISEASE: RESULTS OF LONG-TERM FOLLOW-UP

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Introduction: To assess the mortality rate and determine the causes of mortality in patients with connective tissue diseases (CTDs)-related interstitial lung disease (ILD).

Methods: This study included patients with CTDs-related ILD who were followed up in a tertiary rheumatology outpatient clinic between January 2016 and December 2022.

Results: There were 149 patients diagnosed with CTD-related ILD, and 6 (4.0%) of the patients died during the 5-year follow-up period. The fatality rate was 4.02%. The evaluation of the extent of pulmonary involvement on CT revealed that less than 20% of the parenchyma was affected in 4 patients. According to pulmonary function tests, forced vital capacity was below 70 ml in 3 (50%) patients. All patients received at least one immunosuppressive therapy (IS) for ILD. Four (66.7%) patients were treated with rituximab, 2 (33.3%) with azathioprine, one (16.7%) with mycophenolate mofetil and one (16.7%) with cyclophosphamide. In addition to these treatments, all patients were receiving prednisolone between 7.5 and 30 mg/day. The most important cause of mortality was infections. Five patients died of respiratory failure caused by infectious processes, including 3 patients with COVID-19 infection, one with bacterial pneumonia and one with pneumocystis carinii pneumonia. The other patient with a history of breast cancer died due to cardiopulmonary arrest (Table 1).

Discussion: Our study emphasizes that infections, which are a significant cause of mortality in patients receiving various IS therapies to reduce the progression of interstitial lung disease (ILD) based on the extent or type of pulmonary involvement, warrant increased attention.

Keywords: connective tissue disease, interstitial lung disease, mortality

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6
Age	61	79	64	63	64	57
Gender	Male	Female	Female	Female	Female	Male
CTDs diagnosis	Reumatoid Artritis	Inflammatory Myositis	Inflammatory Myositis	Sjogren's Syndrome	Reumatoid Artritis	Systemic Sclerosis
CTDs duration of illness (months)	382	1	202	16	145	40
Type of ILD pattern	OIP	NSIP	NSIP	NSIP	NSIP	NSIP
ILD disease duration (months)	55	1	202	16	145	40
Immunosuppressive therapy	RTX	AZA	MMF	RTX/AZA	RTX	CYC/RTX
Pulmonary Function Test (FVC<70 ml)	Restrictive pattern	Normal	Normal	Restrictive pattern	Normal	Restrictive pattern
Extent of ILD on CT (>20%)	Yes	No	No	No	Yes	No
Cigarette	Yes	No	No	No	No	Yes
Cause of Death	Infection	Infection	Cardiopulmonary arrest	Infection	Infection	Infection

Table 1. *Demographic characteristics of deceased patients. CTDs: connective tissue disease, ILD: interstitial lung disease, FVC: forced vital capacity, CT: computed tomography, OIP: usual interstitial pneumonia, NSIP: nonspecific interstitial pneumonia, RTX: Rituximab, AZA: Azathioprine, MMF: Mycophenolate mofetil, CYC: Cyclophosphamide*

[Abstract:2817]

FEVER OF UNKNOWN ORIGIN AND SPLENOMEGALY: A CASE REPORT OF VISCERAL LEISHMANIASIS

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Introduction: Visceral leishmaniasis (VL) is a potentially fatal disease endemic to large parts of Africa, Asia, and southeastern Europe. According to 2020 data, 1.13 cases per 100,000 people were reported in Georgia. Visceral leishmaniasis cases were reported in Turkey between 2005–2012 and 2016.

The incidence varies between 0.02 and 0.05 per 100,000 people. The diagnosis remains a challenge due to the non-specific clinical symptoms. Herein we describe a case of fever and splenomegaly of unknown origin, which was finally diagnosed as VL by splenectomy.

A 48-year-old male patient who is considered to have an Adult-Onset Disease in his country Georgia applied to the rheumatology outpatient clinic of our hospital with complaints of fever, sweating and weight loss that had been going on for 3 weeks. In his evaluation, pancytopenia and hepatosplenomegaly were detected. PET/CT was performed, and bone marrow, spleen, and liver involvement was revealed.

However, no diagnostic findings were found in the bone marrow sample. The patient, whose hypersplenism findings deepened and was at risk of progressing to MAS underwent splenectomy for the differential diagnosis of primary splenic lymphoma, MAS associated with Adult-Onset Still's Disease, and visceral leishmania. The pathology evaluation was consistent with visceral leishmania. The patient recovered completely with amphotericin B treatment.

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Early diagnosis and prompt treatment of visceral leishmaniasis are crucial to preventing complications and reduce mortality.

Keywords: *fever, leishmaniasis, splenectomy*

[Abstract:2820]

MEDULLARY GRANULOMATOSIS: UNUSUAL FORM OF SARCOIDOSIS

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Introduction: Diagnosis of sarcoidosis is based on the presence of an epithelial-gigantocellular granuloma without caseating necrosis.

Case Report: a 46-year-old man, on insulinotherapy for type 2 diabetes, was investigated for hypercalcemia. He presented with a dry cough and physical examination revealed grade 1 splenomegaly without palpable lymphadenopathy. Blood counts howed an normocytic normochromic aregenerative anemia (10.5 g/dl of Hb) and thrombocytopenia at 130,000/mm. Creatinine clearance was at 23 ml/min, serum calcium at 146n serum phosphorus at 44 mg/l, calciuria was high and phosphaturia normal. Albuminemia, parathormone Rp and Vit D level were at normal levels, a polyclonal elevation of gammaglobulins, liver tests were normal and angiotensin-converting enzyme level was elevated X2N. The Koch'sb acillus in the sputum was negative, reaction to IDR to tuberculin was anergic and the quantiferon test was negative.

Chest CT revealed multiple bilateral nodular without mediastinal lymphadenopathy. Abdominal CT revealed homogeneous splenomegaly. Imaging of the skeleton was unremarkable.

The cytological and histological study of the bronchial biopsy concluded a non-specific chronic bronchi inflammation, the accessory salivary glands biopsy was unremarkable. Bone marrow biopsy revealed granulomatous lesions made of epithelioid and giant cells without necrosis compatible with sarcoidosis given the clinical context. Cardiac exploration was normal.

The diagnosis of sarcoidosis was retained and corticosteroid therapy was initiated with a quick and spectacular favorable evolution with normalization of biological parameters (calcemia, renal function, anemia).

Conclusions: Sarcoidosis is a diffuse granulomatosis. The medullary localization is a rare hematologic form which represents ≤1.5% of systemic sarcoidosis usually corticosteroid sensitive.

Keywords: *sarcoidosis, bone marrow, medullary sarcoidosis*

[Abstract:2827]

EXPLORING THE PREVALENCE AND ASSOCIATION BETWEEN HYPERURICEMIA AND FATTY LIVER INDEX IN RURAL ROMANIAN COMMUNITIES

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Background: Hyperuricemia is a prevalent condition with potential implications for metabolic health, being a risk factor for nonalcoholic fatty liver disease (NAFLD). Hyperuricemia may also have an impact on the histologic severity of NAFLD. However, data on this association are limited.

Objectives: This study, conducted between March 2023 and November 2023, aimed to investigate the prevalence of hyperuricemia in this population and to determine the association between hyperuricemia and the fatty liver index. The primary objective was to examine the hypothesis that the presence of hyperuricemia is associated with a high fatty liver index.

Methods: Data from 708 unselected participants from rural Romania (500 women and 208 men), with a mean age of 55.62 years was collected and analyzed. Hyperuricemia was defined as serum uric acid > 7.2 mg/dl in men or >6 mg/dl in women. The fatty liver index was calculated using the formula developed by Bedogni et al. NAFLD was defined as fatty liver index >60. The Pearson correlation was employed to assess the relationship between hyperuricemia and the fatty liver index.

Results: There was a high prevalence of hyperuricemia (19.4%) and NAFLD (49%) in the rural population. Hyperuricemia was more often observed in men. A positive and statistically significant Pearson correlation was observed between hyperuricemia and the fatty liver index. This suggests that hyperuricemia has a potential association with the presence of NAFLD as the individuals with hyperuricemia were more likely to exhibit an increased fatty liver index.

Keywords: *epidemiology, hyperuricemia, fatty liver index, population based study*

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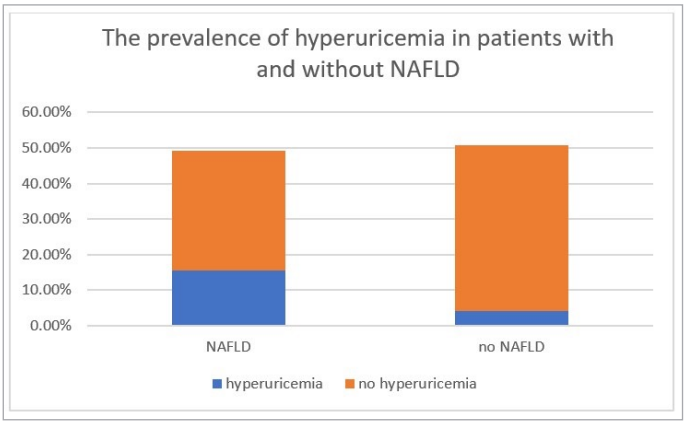


Figure 1. *The prevalence of hyperuricemia in patients with and without NAFLD.*

[Abstract:2879]

CARDIAC MANIFESTATIONS IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction: Systemic lupus erythematosus is a multisystemic disease of unknown etiology. Cardiac involvement remains common and significantly influences the prognosis of the disease. The purpose of our study was to explore its clinical, paraclinical, and therapeutic characteristics.

Patients and Methods: This retrospective descriptive study was conducted in the Internal Medicine Department of Mohamed VI University Hospital in Marrakech, from 2012, to 2022.

Results: Sixty-two patients were included with an average age of 34.57 years and a female-to-male sex ratio of 4.6. Cardiac involvement manifested at the onset in 38 patients. Predominantly, pericardial involvement occurred in 74% of cases. Valvular damage was present in 51.6% of patients. Dilated cardiomyopathy was identified in 27.4% of cases, with an ejection fraction ranging from 15 to 45%. Pulmonary hypertension was observed in 29% of cases, predominantly of moderate severity. Fifty-six patients received methylprednisolone pulse, and 96.7% were on oral steroids with synthetic antimalarials. Seventeen patients with dilated cardiomyopathy were prescribed immunosuppressive therapy for cardiac involvement, primarily using cyclophosphamide. Specific treatment for cardiac damage, including diuretics, anticoagulants, and digitalis, was indicated in 23, 13, and 6 patients, respectively. The outcome was favorable in 83.9% of patients, marked by regression of pericardial effusion in all cases, improvement in ejection fraction in 76.5% of patients with dilated cardiomyopathy, and a decrease in mean pulmonary arterial pressure. However, the outcome was unfavorable in 10 patients, with one patient succumbing to sepsis due to candidemia.

Conclusions: Collaboration between internists and cardiologists is crucial to optimize the care of these patients.

Keywords: *lupus, cardiac manifestations, cyclophosphamid*

[Abstract:2881]

SYSTEMIC LUPUS ERYTHEMATOSUS AND PREGNANCY

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Introduction: A pregnancy associated with lupus poses a high risk of maternal and fetal morbidity and mortality. The objective of our study is to analyze the impact of pregnancy on lupus and illustrate the influence of lupus on pregnancy.

Patients and Methods: This retrospective study examines 54 cases of pregnancy among 357 patients registered in the Internal Medicine Department at Mohammed VI University Hospital, Marrakech, from 2004 to 2022.

Results: The mean age of our patients was 32 years. Out of these, 51 patients were already diagnosed with lupus before their current pregnancy, while lupus was identified during the current pregnancy in three patients.

The average gestational age was 3.5 gestations, and the average parity was 2.1 parities. Intrauterine growth restriction and pre-eclampsia was observed at 8.33%. Prematurity was observed in 4.17%. The clinical manifestation of lupus primarily involved osteoarticular and mucocutaneous aspects (90%). Lupus glomerulonephritis was present in 40% of cases, predominantly class IV, cardiac involvement in 43%, and neuropsychiatric involvement in 33%. Partial remission during pregnancy was noted in 30% of patients, with a lupus relapse in 10%. Postpartum relapses were experienced by 44% of patients.

Treatment included systemic steroids (89%), synthetic antimalarials (70%), and azathioprine (20.7%). Antiplatelet agents were prescribed in 42% of cases, while heparin was administered in 12%. Antihypertensive drugs were used in 12% of cases.

Conclusions: Pregnancy in women with lupus should involve preconception consultation and be planned during a period of at least 6 months to minimize the risk of relapses and obstetric complications.

Keywords: *pregnancy, lupus, steroids*

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[Abstract:2883]

OCULAR INVOLVEMENT IN BEHÇET'S DISEASE: AN ANALYSIS OF 83 PATIENTS (149 EYES)

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Introduction: Behçet's disease is a chronic, multi-system vasculitis of unknown etiology. The eye is frequently affected. Uveitis and retinal vasculitis are the major manifestations of the disease. The aim of our study is to describe the clinical and evolution modalities of ocular involvement in Behçet's disease, in our department.

Patients and Methods: This is a retrospective study of patients with ocular involvement of Behçet's disease, seen from January 2010 to December 2022 in the internal medicine department of the university hospital Mohammed VI, Marrakech. All patients met the classification criteria of the International Study Group for Behçet's Disease.

Results: 83 patients were enrolled with a male/female ratio of 2.2. Ophthalmic manifestations were the initial presentation of the disease in 67% of patients. The most common ocular findings were retinal vasculitis (48%) and intermediate uveitis (42%). Main drug therapy consisted of colchicine, steroids, immunosuppressive therapy (cyclophosphamide and azathioprine) and biologic agents (TNF inhibitors and tocilizumab). The most important complications were synechiae (28%), blindness (16%) and macular edema (14%).

Conclusions: Ocular involvement in Behçet's disease is a serious condition, and can lead to blindness. A close collaboration between the ophthalmologist and internist is required to improve the prognosis.

Keywords: *Behçet, uveitis, retinal vasculitis*

[Abstract:2893]

SYSTEMIC LUPUS ERYTHEMATOSUS: A RARE FINDING LEADING TO DIAGNOSIS

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Case Description: 25 year-old woman presented to our clinic for a second opinion diagnosis, with a previous two year history of painful subcutaneous nodules on her arms, trunk, neck and inguinal area, that she was managing with traditional chinese medicine. She had previously performed a cutaneous biopsy that showed a hypodermis lymphocytic infiltrate predominantly surrounding adipocytes, with irregular nucleus and some in karyorrhexis, suggesting the hypothesis of subcutaneous panniculitis-Like T-cell lymphoma (SPTCL). Further investigation revealed no significant results and a new cutaneous biopsy was performed showing a perivascular inflammatory infiltrate in dermis and hypodermis, with small lymphocytes, with no atypical features and no T cell expression anomalies. No granulomas or vasculitis were seen. At this moment, the patient was diagnosed with LEP.

Clinical Hypothesis: Lupus erythematosus panniculitis (LEP) versus SPTCL.

Diagnostic Pathways: Repeated follow-up blood tests showed mild thrombocytopenia, leukopenia, ANAs of 1/320, Anti-DsDNA elevation and mild C3 consumption. Full-body CT-scan showed multiple ganglionar formations, without signs of suspicion. New cutaneous biopsy was compatible with LEP, with normal immunohistochemistry, immunophenotyping of T cells and no clonal TCR gene rearrangements. Given the findings, systemic lupus erythematosus with cutaneous and hematological involvement was made, and treatment with prednisolone, hydroxychloroquine and azathioprine was initiated, with significant improvement of lesions.

Discussion and Learning Points: Both these conditions are rare and clinically very similar. Histologic findings can help differentiate them, but there are cases of overlapping features. Immunophenotyping, immunohistochemistry and evaluation of TCR gene rearrangements are fundamental to achieve diagnosis and define the appropriate approach.

Keywords: *lupus, panniculitis, T-cell lymphoma*

[Abstract:2910]

DYSPNEA

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A 65-year-old woman with a history of hypertension, diabetes and chronic kidney disease consulted for dyspnoea, fever and asthenia. On arrival, febrile, haemodynamically stable although tachypnoea. Auscultation with bibasal crackles and presence of generalised oedema.

Laboratory tests showed leukocytosis, normocytic anaemia, creatinine 8.36 mg/dL, C-reactive protein 372 mg/L, procalcitonin 4 ng/mL and NT-proBNP at 8700 pg/mL. Chest X-ray showed bilateral alveolar infiltrates and ultrasound showed kidneys with a chronic appearance. Serology for atypical pneumonia and blood cultures were requested. Empirical antibiotic therapy was started on suspicion of sepsis of respiratory origin.

Given the volume overload and oligoanuria, admission to the Intensive Care Unit for haemodiafiltration was decided. Initial evolution was poor despite antibiotic therapy, with progressive anaemia requiring multiple transfusions, worsening pulmonary infiltrates and no improvement in renal function.

In view of the diagnostic uncertainty and in the absence of

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response to treatment, the patient was referred to Internal Medicine. Considering the reno-pulmonary involvement autoimmune test and fibrobronchoscopy were requested. In this study, ANCA-MPO antibodies > 143 IU/ml and hypoproteinaemia, with albumin/creatinine ratio 4042 mg/g were found. In addition, haemosiderophages in bronchoalveolar lavage.

Based on the presence of nephrotic syndrome and alveolar haemorrhage with the analytical results, the diagnosis of vasculitis due to anti-myeloperoxidase antibodies is assumed. Considering the respiratory involvement and the aspect of renal chronicity, it was decided not to perform a biopsy, and treatment was started with corticoids, plasmapheresis and cyclophosphamide, obtaining a favourable evolution with complete remission of the condition except for renal function, currently requiring haemodialysis.

Keywords: *fibrobronchoscopy, haemodiafiltration, haemosiderophages, anti-myeloperoxidase antibodies, vasculitis*



Figure 1. *Axial non-contrast chest CT reconstruction.*

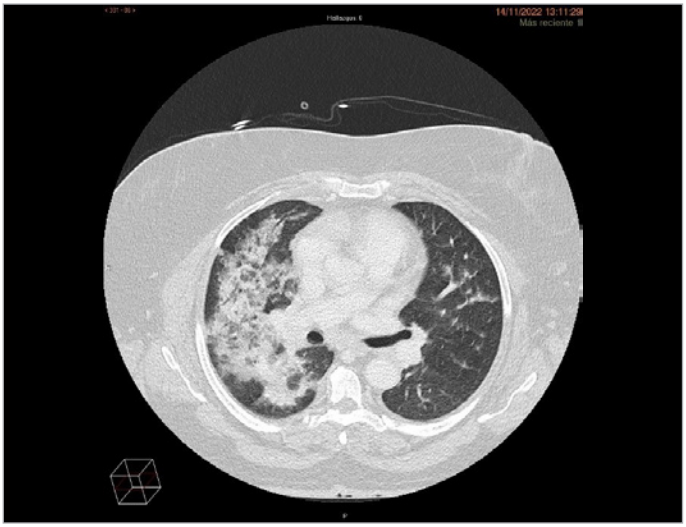


Figure 2. *Coronal non-contrast chest CT reconstruction.*

[Abstract:2911]

AN UNCOMMON CASE OF RITUXIMAB INDUCED BICYTOPENIA

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Case Description: A 72-year-old man was admitted to the hospital with a one-week history of new onset hemoptysis and hematuria. He had a history of COPD, vitiligo, and hypertension. Upon physical examination, he appeared apyretic and had diminished breath sounds. In his lab results 7.4 g/dl of hemoglobin, 153,000 platelets per milliliter, 8.75 white blood cells per milliliter, 5.75 creatinine, MPO-ANCA >200, and P-ANCA++.

The kidney biopsy revealed crescentic glomerulonephritis and the patient was diagnosed with microscopic polyangiitis. Rituximab (RTX), isoniazid prophylaxis, and 80 mg of methylprednisolone were administered. One gram of RTX (1g in every 15 days/6 months) was started as the first dose and one month later 500 mg RTX was administered. PLT and WBC levels were 96000 10 3/uL and 3.43 10 3/uL, respectively, following the second dose. Based on our investigation into the etiology of recently developed bicytopenia, it is believed to have been caused by rtx and was subsequently stopped.

Clinical Hypothesis: Peripheral blood cytopenia after rtx injection

Diagnostic Pathways: Blood tests, kidney biopsy, computer tomography, urine tests.

Discussion and Learning Points: Even if bicytopenia is not a frequent adverse effect of rtx, it may be important to remember that rtx may be connected to newly developed bicytopenia following an injection. After 45 days after stopping the medication, the bicytopenia subsided.

Keywords: *rituximab, bicytopenia, rtx, hemoptysis, hematuria*

[Abstract:2922]

THYRO-GASTRIC AUTOIMMUNE SYNDROME

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Introduction: Thyro-Gastric Autoimmune Syndrome (TGAS), firstly described in the 1960s, is the association of two autoimmune diseases including thyroid involvement (Hashimoto's thyroiditis or Graves' disease) and gastric mucosa involvement (pernicious anemia, an autoimmune disease affecting the stomach).

Methods: This is a descriptive retrospective study conducted over

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18 years. We included 8 patients followed for TGAS in the internal medicine department in Sahloul University Hospital.

Results: Pernicious anemia (PA) or Biermer disease associated with thyroid dysfunction was found in 5 women and 3 men. The average age at diagnosis was 57.75 years, ranging from 37 to 77 years. Hashimoto's thyroiditis was found in 6 cases (75%) and Graves 'disease in 2 cases (25%). PA preceded autoimmune thyroiditis in 3 patients (37.5%).

Macrocytic anemia was observed in 7 cases (87.5%) with an average hemoglobin level of 8.76 g/dL. Anti-parietal cell antibodies were present in 3 patients (37.5%), while anti-intrinsic factor antibodies were positive in only 5 patients (62.5%). Digestive endoscopy was performed in 6 patients (75%), revealing villous atrophy in 2 cases (25%). The treatment for PA consisted of lifelong intramuscular vitamin B12 injections, with favorable clinical and biological outcomes in all 8 patients.

Conclusions: Thyro-Gastric Autoimmune Syndrome represents a unique example of the complexity of autoimmune diseases. Some studies emphasize genetic and hereditary susceptibility, while environmental aspects, particularly infectious factors in gastritis and autoimmune thyroiditis, are often overlooked.

Keywords: *pernicious anemia, Hashimoto's thyroiditis, Graves 'disease*

[Abstract:2923]

ASSESSING THE INFLUENCE OF DIVERSE BIOLOGICAL AND TARGETED SYNTHETIC THERAPIES ON TOTAL CHOLESTEROL LEVELS IN RHEUMATOID ARTHRITIS: A SCIENTIFIC EXPLORATION

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Background and Aims: The objective of this study was to elucidate changes in total cholesterol values after the initiation of biological therapy compared to previous values. Biological and targeted synthetic therapy in rheumatoid arthritis may influence total cholesterol.

Methods: We conducted a retrospective study involving patients with rheumatoid arthritis undergoing targeted biological and synthetic treatment.

Among them, 24.39% were treated with JAK-inhibitors, 20.32% with rituximab, 20.32% with adalimumab, 20.32% with etanercept and 14.63% with tocilizumab. Inclusion criteria comprised paraclinical assessment of total cholesterol before initiation of biologic therapy and at least two subsequent follow-up assessments. The paired T-test was employed to compare total cholesterol values before and after the initiation of the specific treatment.

Results: A total of 123 patients with rheumatoid arthritis were included. For the entire group, total cholesterol values increased on average by 11.66 mg%, CI 95% [17.47-5.85] mg%, p<0.001. In the case of treatment with JAK-inhibitors, total cholesterol values increased on average by 19.18 mg%, CI 95% [29.70-8.65] mg%, p=0.001, and by 12.21 mg% on average in those undergoing rituximab treatment, CI 95% [20.86-3.56] mg%, p=0.008. Patients treated with adalimumab, etanercept and tocilizumab showed statistically insignificant increases in values.

Conclusions: The study identified a statistically significant increase in total cholesterol values in the entire group, as well as in patients treated with JAK-inhibitors and rituximab. This suggests that the impact of biological and targeted synthetic therapies on the lipid profile requires special attention in monitoring this phenomenon.

Keywords: *rheumatoid arthritis, biological therapy, targeted synthetic therapy, total cholesterol*

[Abstract:2926]

IN THERAPEUTIC PRACTICE, PATIENTS WITH THE ADULT ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) AND LOW DISEASE ACTIVITY HAVE A LOWER QUALITY OF LIFE AS COMPARED WITH ADULT PATIENTS WITH JUVENILE SLE

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Purpose: To compare the quality of life (QoL) among the adult patients (pts) with low activity of SLE and juvenile (<18 years) and adulthood (≥18 years) onset of the disease.

Methods: Totally, 68 pts with SLE and low disease activity (SLEDAI-2K≤4) were involved: 35 pts with juvenile SLE (jSLE) and 33 pts with adult SLE (aSLE). The QoL was assessed by MOS SF-36 (Short form Medical Outcomes Study) questionnaire.

Findings: Mean age of pts with jSLE and aSLE was 29.8±8.05 and 38.3±7.69 years, disease duration was 5 [1; 25] and 3 years [1; 21] respectively (p<0.05 for all). The average daily dosage of glucocorticoids in prednisolone was 7.57±5.13 mg and 9.1±5.5 mg/day respectively (p=0.05), 37% of patients with jSLE and 27% with aSLE received cyclophosphamide (p≥0.05). jSLE pts had a higher MOS SF-36 mental health score compared with aSLE pts (55.4 ± 7.26 vs. 50.7 ± 8.7, p = 0.012). In patients with aSLE a correlation between the daily dosage of glucocorticoids, the frequency of cyclophosphamide usage and with role-physical functioning was found, r=0.51, p<0.05.

Conclusions: Patients with low activity of SLE have higher level of the mental health problems in aSLE as compared with jSLE patients.

The role of physical functioning in daily activities in aSLE was

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interrelated with higher dosage of glucocorticoids and frequent administration of the cyclophosphamide.

Keywords: *quality of life, juvenile-onset systemic lupus erythematosus, low disease activity*

[Abstract:2927]

MULTIPLE AUTO-IMMUNE SYNDROME

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Introduction: Multiple Autoimmune Syndrome (MAS) is the association of at least three distinct autoimmune diseases (ADs).

Methods: This is a descriptive retrospective study conducted over a 19-years period (2004-2023) in the internal medicine department in Sahloul University Hospital, collecting data from patients with MAS.

Results: The study included 19 patients, comprising 16 females and 3 males, with a female-to-male ratio of 5.3. The average age at the diagnosis of the first autoimmune pathology was 43.7 years [19-63 years]. MAS involved three ADs in 16 patients, four in one patient, five in one patient, and six ADs in another patient.

According to the Humbert and Dupond classification of ADs, one patient was classified in MAS type I, one in MAS type II, and 7 in MAS type III. These ADs were distributed as follows: systemic lupus erythematosus was the most frequently observed (13 cases, 68%), followed by Sjogren's syndrome (63%, n=12), autoimmune thyroiditis (36%,n=7), vitiligo (36%,n=7), rheumatoid arthritis (15%, n=3), systemic sclerosis (15%, n=3), autoimmune haemolytic anemia (15%, n=3),primary biliary cirrhosis (15%, n=3), dermatomyositis (10%, n=2), pernicious anemia (5%, n=1), thymoma (5%, n=1), type 1 diabetes (5%, n=1), adrenal insufficiency (5%, n=1) and one case of psoriasis. Other associated ADs not belonging to the three types of MAS described were found: celiac disease in 4 patients and autoimmune hepatitis in two patients.

Conclusions: Multiple Autoimmune Syndrome represents a significant medical challenge for both patients and healthcare professionals.

Keywords: *multiple autoimmune syndrome, autoimmune diseases, descriptive retrospective study*

[Abstract:2931]

FORTUITOUS DISCOVERY OF SEGMENTAL ARTERIAL MEDIOLYSIS SYNDROME

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Introduction: Segmental Arterial Mediolysis (SAM) is a rare, non-atherosclerotic, and non-inflammatory disease of unknown etiology that affects medium-sized arteries.

We report a case of SAM incidentally discovered through imaging in an asymptomatic adult male.

Case: A 54-year-old man, with a 30-pack-year smoking history, was under urological follow-up for obstructive right renal pelvis stone with an indication for pyelotomy. Preoperative imaging in November 2023 revealed a peri-vascular tissue sleeve reducing arterial calibers encompassing the celiac trunk and its branching arteries, associated with splenic infarction. He was admitted to our internal medicine service for further investigation, although the patient had no complaints.

Upon admission, the clinical examination was normal. Laboratory tests were strictly normal: no biological inflammatory syndrome, normal lipid profile, HbA1C% at 5.3%, normal plasma protein electrophoresis, and negative immunological tests (ANCA, AAN, and anti-phospholipid antibodies). Infectious workup was negative.

An aortic and branch angiogram performed in our department two months after the first scan showed a marked regression of the peri-vascular tissue sleeve around the celiac trunk, common hepatic artery, and splenic artery, partial regression of the splenic infarct, and no newly appeared vascular lesions.

The diagnosis of SAM was established after ruling out other potential causes: bacterial or fungal infections, vasculitis, and non-inflammatory. The predominantly splanchnic circulation involvement, absence of associated systemic signs, and especially the spontaneous regression of the tissue sleeve were crucial in making the diagnosis.

Conclusions: This case describes an incidentally discovered asymptomatic case of SAM with spontaneous regression.

Keywords: *asymptomatic, segmental arterial mediolysis, spontaneous regression*

[Abstract:2933]

DIFFUSE INTERSTITIAL LUNG DISEASES

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Introduction: Diffuse Interstitial Lung Diseases (ILDs) represent a heterogeneous group of pulmonary disorders characterized by

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inflammation and fibrosis affecting the lung interstitial tissue. The etiologies of ILDs are varied.

Methods: A descriptive retrospective study conducted over an 18-year period. We included 24 patients presenting with ILD at the internal medicine department at Sahloul University Hospital.

Results: We collected data from 24 patients. There were 11 men and 13 women, with a male-to-female ratio of 0.846. The average age at the time of ILD diagnosis was 53 years (range: 25 to 80 years). Chest X-ray showed interstitial syndrome in 10 cases (41%) and was normal in 14 cases (58%). Thoracic computed tomography revealed subpleural reticulations in 5 cases (20%), micronodules in 17 cases (70%), ground-glass opacities in 3 cases (12%), honeycombing in 2 cases (8%), and peri-broncho-vascular thickening in 5 cases (20%).

ILD etiologies were dominated by sarcoidosis in 8 cases (33%), systemic sclerosis in 3 cases (12%), Sjogren's syndrome in 2 cases (8%), rheumatoid arthritis in 2 cases (8%), inflammatory myopathy in 2 cases (8%) (dermatomyositis), common variable immunodeficiency in 2 cases (8%), pulmonary tuberculosis in 2 cases (8%), Kikuchi-Fujimoto disease, Erdheim-Chester disease, and pulmonary involvement of lymphoma, each in one case (4%).

Aggravation was observed in 2 patients (8%), with one case of chronic respiratory failure (4%).

Conclusions: The etiologies of diffuse interstitial lung diseases are multiple and complex, involving intricate interactions between genetic, environmental, and immunological factors.

Keywords: diffuse interstitial lung diseases, descriptive retrospective study, etiologies

[Abstract:2962]

AN EFFECTIVE THERAPEUTIC TREATMENT OF CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO) IN ADULTS. A SERIES OF 11 CLINICAL CASES

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Summary: The data of the course and therapeutic options of CRMO as an orphan bone disease in adults are limited [1].

Purpose was to investigate the features and possibilities of treatment of CRMO in adults.

Patients and Methods: A sample of 11 histologically verified cases was formed from the 1650 patients, follow-up period was 1 year.

Findings: The characteristics of the patients are presented in Table 1.

Conclusions: CRMO in adults have no differences with paediatric CRMO. Drug treatment of CRMO in adults is more effective than surgical treatment.

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Keywords: chronic, recurrent, multifocal, osteomyelitis

	Mean age, years	Clinical manifestations, years	X-ray detected osteitis, years
Male (n=5)	38.99±17.45	29.2±19.08	31.00±18.29
Female (n=6)	35.96±14.23	11.5±6.59	14.0±9.67
Total	35.7±15.26	21.33±16.56	23.44±16.53

Table 1. Age of osteitis manifestation, n=11.

Drugs	Patients, n (%)
NSAIDs	11 (100)
Glucocorticoids, per os	3 (27.27)
Sulfasalazine, 2–3 g/day	4 (36.36)
Methotrexate, 15–25 mg/week	1 (9.09)
Mycophenolate mofetil, 1-3 g/day	1 (9.09)
Leflunomide, 20 mg/day	1 (9.09)
Zoledronic acid, 5 mg	2 (18.18)
Pamidronic acid, 90 mg	1 (9.09)
Adalimumab, 40–80 mg/2 weeks	2 (18.18)
Infliximab, 5 mg/kg / 8 weeks	2 (18.18)
Golimumab, 50 mg/4 weeks	1 (9.09)

Table 2. Drug treatment, n=11.

Surgery	Patients, n (%)
Bone resections	6 (100)
Relapses after bone resection	6 (100)
Surgery on immunosuppressive therapy	0 (0)

Table 3. Surgical treatment, n=6.

[Abstract:2969]

CHEST PAIN AND ANTIPHOSPHOLIPID SYNDROME (APS). AN ASSOCIATION THAT SHOULD NEVER BE IGNORED

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Case Description: A 46-year-old woman, with mild anxiety, smoker, with chronic hypertension headache, systemic lupus erythematosus (SLE), triple positive for antiphospholipid antibodies on treatment with hydroxychloroquine since the

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beginning of 2023 and episodes of typical chest pain since the beginning of 2022 with Mibi-spect, coronary CT, CT angiography and echocardiogram with normal results prior to admission. She presented with a new episode of transfixing chest pain of few seconds of duration, associated with diaphoresis, skin pallor and not associated with physical stress.

Clinical Hypothesis: Causes of chest pain in young woman with history of anxiety, classic cardiovascular risk factors and negative ischemia tests.

Diagnostic: Patient persists with chest pain during admission. An EKG during chest pain showed negative T waves in V1-V4. Stress test (MIBI-SPECT) was positive, showing ST elevation (2 mm) in lower precordials. Cardiac catheterization concluded a severe stenosis (80%) in the middle segment of the LAD that required a stent.

Discussion and Learning Points: Due to the pathologic background and the coronary event an APS was considered and anticoagulation treatment was started. APS is an autoimmune disease, characterized by the presence of antiphospholipid antibodies, which can cause arterial and venous thrombosis. It affects coronary arteries in up to 2.8% of cases. So, in this patient even in the absence of a typical presentation, and positive test results and in a scenario of an anxiety component, we should not forget that, especially in the presence of risk factors, APS might be considered as a differential diagnosis.

Keywords: *SLE, APS, chest pain*

[Abstract:3019]

ANCA-ASSOCIATED VASCULITIS PRESENTING AS RENAL-LIMITED VASCULITIS: A CASE REPORT

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Case Description: We report the case of a 39-year-old woman from Guinea-Bissau, with a medical history of cured hepatitis B virus (HBV) infection. She presented to our hospital with vomiting and abdominal pain, having previously been treated with cefixime for a urinary tract infection. Due to progressive worsening of renal function in the last month with nephrotic proteinuria, she was admitted for further investigation.

Clinical Hypothesis: Systemic autoimmune disease, HBV reactivation, multiple myeloma, and acute interstitial nephritis secondary to antibiotic therapy.

Diagnostic Pathways: She presented anemia, ESR 129 mm/1h; no complement consumption, ANAs of 1/640 and positivity for ANCA-MPO, with remaining autoimmune study being negative. Viral serologies were negative, including HBV viral load. Proteinogram and measurement of light chains were normal. Full-body CT scan revealed no significant findings. Renal biopsy showed 13 glomeruli, 4 cellular crescents with fibrinoid necrosis, a lymphocytic inflammatory infiltrate, without granulomas. Immunofluorescence showed only slight deposition of C3c, with no other deposits, compatible with ANCA-MPO vasculitis.

Therefore, treatment was initiated with pulses of methylprednisolone and rituximab associated with entecavir as prophylaxis. A significant improvement in renal function and proteinuria was seen two weeks after.

Discussion and Learning Points: ANCA-vasculitis are rare diseases, with renal-limited vasculitis (RLV) being even rarer. Both have an incidence typically around the age of 60, making our case even less common. Despite RLV having better outcomes in terms of recurrence, the need for dialysis or renal transplantation, it still carries a 90% mortality rate in untreated patients. Therefore, prompt diagnosis and aggressive therapy are crucial.

Keywords: *vasculitis, ANCA-MPO, renal-limited*

[Abstract:3062]

ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA WITH PANCREATITIS AT DISEASE ONSET IN SYSTEMIC LUPUS ERYTHEMATOSUS: SUCCESSFUL MANAGEMENT OF A RARE PRESENTATION

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Systemic lupus erythematosus (SLE) is a complex autoimmune disease. Any type of organ involvement could be found in patients with SLE.A cute pancreatitis as the initial manifestation of SLE has been rarely reported. Mechanisms of pancreatitis in SLE patients include immune complexes precipitation in the pancreatic vessels, micro thrombosis, antiphospholipid syndrome or drugs such as tacrolimus, azathioprine. Thrombotic thrombocytopenic purpura (TTP) in patients with SLE is extremely rare. TTP as the first manifestation of SLE is even rarer. Here, we report a patient presented with acute pancreatitis and diagnosed with SLE related TTP.

The case is a 57-year-old woman without any history of chronic disease referred to emergency department with fever, stomach ache and confusion. She had a tenderness of epigastric region. Laboratory results are shown in table 1. Complement C3 and C4 were low. Antinuclear antibody (ANA) was 1/1280 titer. Plasma exchange and corticosteroid treatment were started urgently for the treatment. In follow-up patients platelet count increased and also her confusion improved. A disintegrin and metalloproteinase

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with thrombospondin type 1 motifs, member 13 (ADAMTs-13) activity was detected 0.25%, antigen level was in normal ranges but the inhibitor level was determined high. She had proteinuria (3g/day) and hematuria on microscopic assay. The diagnosis of SLE based on the Systemic Lupus International Collaborating Clinics (SLICC) criteria with high ANA concentration, low complement, proteinuria and neurologic involvement was established. Cyclophosphamide was given as maintenance therapy after plasmapheresis due to TTP associated with SLE.

In our patient, it took a high index of suspicion spurred by the emergence of low complement assay, an immunologic finding, proteinuria and neurologic involvement. Awareness should be given to the possible autoimmune diseases in acute pancreatitis.

Keywords: *TTP, pancreatitis, thrombotic microangyopathy, systemic lupus erythematosus*

Values \ Treatment		PE – D1	PE – D2	PE – D3	PE – D4	PE – D5	PE – D6
					750 MP	750 MP	750 MP
Urea (mg/dL)	219	103	106	73	56	62	59
Creatinin (mg/dL)	2.0	1.3	1.0	0.8	0.7	0.7	0.7
TB/CB (mg/dL)	2.1/1.4	5.7/4.8	3.1/2.1	4.2/3.0	2.0/1.2	1.7/1.08	1.4/0.7
LDH (U/L)	1095	858	477	572	474		372
Haptoglobin (mg/dL)	<10	<10	<10	<10			
Reticulocytes, %	16.3	13.6	9.7	9.3	9.2	8.7	8.6
Platelet (10^{x3}/µL)	26.000	6.000	19.000	10.000	55.000	117.000	166.000

PE, plasma exchange; D,day; MP, methylprednisolone; TB, total bilirubin; CB, conjugated bilirubin; LDH,lactate dehydrogenase.

Table 1. *Laboratory results and treatment data*

[Abstract:3073]

ANTISYNTHETASE SYNDROME: A MISDIAGNOSED CASE PRESENTING WITH MYOCARDITIS IN INTENSIVE CARE UNIT

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Introduction and Aim: Antisynthetase Syndrome (ASS) is an autoimmune disease characterized by the triad of arthritis, myositis and interstitial lung disease (ILD). The diagnosis can commonly be confused with Rheumatoid Arthritis (RA). We report a case ASS complicating with myocarditis.

Case: 49-year-old woman who had been previously diagnosed with RA admitted to the emergency service with shortness of breath, edema in lower extremities, weakness in proximal muscles and dysphagia. The patient was admitted to internal medicine intensive care unit with suspected myocarditis due to bilateral pleural effusion, increased bilateral interstitial markings on her chest X-ray and troponin T level of 800 ng/L. The patient was dyspneic, hypotensive and tachycardic, muscle strength was 2/5. Laboratory results revealed increased CK, troponin T, NT-proBNP, ferritin and CRP (Table 1). Anti-CCP titer was normal. ANA titer was granular pattern 1/1280. Anti-Jo 1 test, which was requested due to proximal muscle weakness and high CK results was positive, also measured by ELISA was >200 RU/mL. When her previous records were analyzed, it was seen that elevated levels of CK were also present during the period when she was diagnosed with RA. Pulse steroid and cyclophosphamide treatment was started with the diagnosis of ASS associated myocarditis due to EF being 56% and elevated levels of CRP, ferritin, NT-proBNP and troponin. Further tests concerning any possible malignancies and viral etiologies revealed negative. She was discharged on the 15th day of treatment with clinical improvement.

Conclusions: Myocarditis is a rare but potentially life-threatening manifestation of some auto-immune diseases. Diagnosis is usually made based on clinical presentation, non-invasive imaging findings, classically echocardiography. Glucocorticosteroids and immunosuppressive agents remain the cornerstone of the treatment. This case report implies interesting considerations about inflammatory cardiac involvement in an autoimmune disease.

Keywords: *antisynthetase syndrome, myocarditis, myositis*

	August 2021	September 2021	October 2021	Before Treatment	15th day of treatment	Normal range
Creatine kinase	2197 IU/L	786 IU/L	801 U/L	306 IU/L	120 IU/L	34-145 IU/L
Troponin T	-	806 ng/L	820 ng/L	487 ng/L	269 ng/L	<14 ng/L
NT-proBNP	-		892 ng/L	5360 ng/L	1009 ng/L	<125 ng/L
CRP	15.8 mg/L	190 mg/L	15.5 mg/L	16 mg/L	3.56 mg/L	0-5 mg/L
Ferritin			1016.2 µg/L	693 µg/L	430 µg/L	13-150 µg/L

Table 1. *Laboratory results.*

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