EJCRIM
European Journal
of Case Reports
in Internal Medicine

POLYMYALGIA RHEUMATICA DURING THE COURSE OF LIMITED CUTANEOUS SCLERODERMA: AN UNUSUAL AND CHALLENGING COEXISTENCE

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Received: 14/06/2024
Accepted: 17/06/2024
Published: 01/07/2024

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent was obtained from patient.

How to cite this article: Lorenzo-Villalba N, Habib C, Docquier L, Nasco E, Imperiale A, Andrès E, Terrade JE. Polymyalgia rheumatica during the course of limited cutaneous scleroderma: an unusual and challenging coexistence. *EJCRIM* 2024;**11**:doi:10.12890/2024_004709

ABSTRACT

A 50-year-old patient with a history of limited cutaneous scleroderma began with polyarthralgia (left shoulder, elbows and hips) without stiffness or associated inflammatory syndrome. Treatment with oral anti-inflammatory drugs was started on suspicion of peripheral spondyloarthritis with partial response. This progressed with the appearance of stiffness and functional limitation of the hips as well as an increase in the inflammatory syndrome two weeks after onset. It was decided to perform an ^{18}F-FDG-PET scan compatible with polymyalgia rheumatica. The patient was treated with oral corticosteroids with an excellent response after one week of treatment.

KEYWORDS

Polymyalgia rheumatica, scleroderma

LEARNING POINTS

- Polymyalgia rheumatica should be considered, even in young adults, with atypical clinical presentation.
- Post-infectious and paraneoplastic inflammatory rheumatism should be ruled out before considering the diagnosis of polymyalgia rheumatica.
- ^{18}F-FDG-PET plays an important role in the positive diagnosis of PMR and in the differential diagnosis.

INTRODUCTION

The prevalence of scleroderma has been estimated to be between 4 and 253 cases per million[1]. Polymyalgia rheumatica (PMR) is a relatively common disease in European patients older than 50 years[2,3] and its prevalence is 700 cases per 100,000 population[1]. However, the prevalence of these conditions occurring together is unknown and scarce reports can be found. The diagnosis of PMR could be challenging in the course of scleroderma.

CASE DESCRIPTION

A 50-year-old woman was admitted to the internal medicine

department for reassessment of polyarthralgia of the left shoulder, elbows and hips, which had progressed for about two weeks without improvement on anti-inflammatory therapy. The patient had a history of limited scleroderma diagnosed in 2019, hypothyroidism diagnosed in 2006, and chronic low back and neck pain with a herniated L5-S1 disc operated on in 2001. Her family history was relevant for rheumatoid arthritis and indeterminate polyarthritis in her mother, who was on treatment with levothyroxine 100 µg daily.

The patient presented with inflammatory polyarthralgia of the left shoulder and hip, following which she was referred to internal medicine. X-rays of the shoulders and hips revealed no abnormalities, and an ultrasound scan of the hips revealed tendonitis at the insertion of the femoral muscles bilaterally. Biological tests revealed no inflammatory syndrome (leukocytes, C-reactive protein and erythrocyte sedimentation rate), so shortly after discussion with our rheumatologist colleagues, she started treatment with celecoxib 100 mg ×2 daily, which was increased to 200 mg ×2 daily 10 days later in view of an incomplete response to treatment. In the meantime, the patient again underwent laboratory tests, which revealed no inflammatory syndromes. It should be noted that she had presented a similar episode of short duration in 2021 with resolution under analgesics, and at that time she had benefited from an magnetic resonance imaging (MRI) of the sacroiliac and cervical, dorsal and lumbar spine, which did not find any element in favour of a spondyloarthritis, but this examination described degenerative disc disease and florid posterior articular osteoarthritis (L4–L5 level) causing severe canal narrowing (Schizas D) and bilateral foraminal narrowing.

On admission to the internal medicine ward the patient was normotensive, heart rate was 80 bpm and she was apyretic. Heart sounds were regular; no murmurs, no oedema and no signs of deep venous thrombosis were found. There were no skin lesions other than facial telangiectasia and skin sclerosis limiting mouth opening. No limb ulcers were found, and no arthritis was noted on clinical examination.

The blood test revealed C-reactive protein of 29.1 mg/l, and erythrocyte sedimentation rate was 40 mm without hyperleukocytosis; normocytic anaemia was 10.7 g/dl with a corpuscular volume of 69 Fl and no thrombocytosis. Liver and renal function tests were unremarkable. Creatine kinase was within normal limits, and cortisol and thyroid function were within normal range. Protein electrophoresis showed no abnormalities. Serology for *Coxiella burnetii*, cytomegalovirus, Epstein-Barr, parvovirus B19, HIV, hepatitis B virus, hepatitis C virus and syphilis was negative. A polymerase chain reaction test for parvovirus B19, and urine tests for gonococcus and chlamydia were negative.

The autoimmune work-up revealed positive antinuclear antibodies at a titre greater than 1/1280, with the presence of anti-centromere antibodies. Anti-RNP, anti-Scl-70, anti-Ro and anti-La antibodies were negative, as were anti-Mi-2, anti-TIF-1 gamma, anti-SAE 1 and 2, anti-NXP-2, anti-MDA-5, anti-Jo-1, anti-PL-7 and 12, EJ, OJ, KS, Zo, Ha and anti-HMGCR. Anti-SRP, anti-DNA, anti-CCP and rheumatoid factors were negative, as were p-ANCA and c-ANCA. Complement was within the standard. The patient benefitted from a knee echography, which showed no joint effusion.

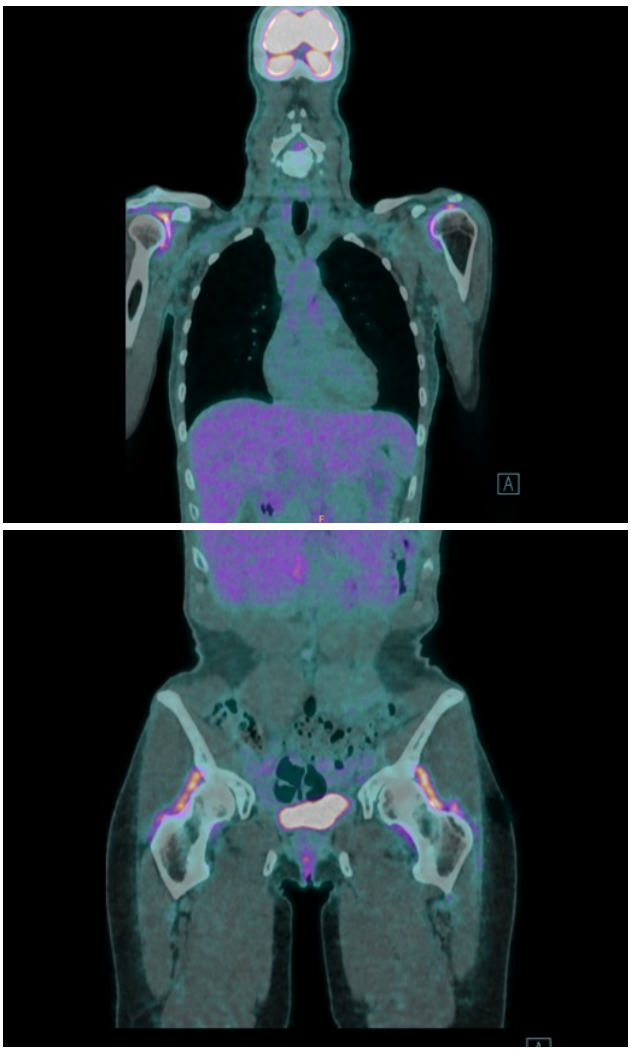

Figure 1. ^{18}F-FDG-PET scan.

The initial hypothesis of peripheral spondylitis was ruled out in view of the partial response to treatment with non-steroidal anti-inflammatory drugs, which was suspended. In addition, at the time of hospitalisation, morning stiffness lasting more than 45 minutes was observed, with an increase in the inflammatory syndrome. In this context, we suggested a paraneoplastic inflammatory polyarticular syndrome or even vasculitis of the large vessels, and an ^{18}F-FDG-PET scan was performed. This showed diffuse periarticular hypermetabolism of the shoulders and hips, bilateral hypermetabolism of ischial tendon insertions suggestive of proximal hamstring tendinopathy, C5 to C7 and L2 to L3 interspinous hypermetabolism suggestive of cervical and lumbar interspinous bursitis, and more discrete hypermetabolism of bilateral sternoclavicular joints. These features were consistent with polymyalgia rheumatica *(Fig. 1)*.

The patient was therefore started on prednisone 15 mg/day, combined with calcium and vitamin D potassium supplementation and gastric protection with omeprazole 20 mg. Clinical evolution was rapidly favourable after seven days of treatment, with improvement of the pain syndrome and functional limitation, and a biological reduction in the inflammatory syndrome.

DISCUSSION

Systemic sclerosis (SSc) and PMR are uncommon but not rare rheumatic diseases. The various musculoskeletal symptoms presenting in SSc can make the recognition of PMR challenging in these patients[4]. Musculoskeletal manifestations are frequent in scleroderma and may be the initial presentation of the disease as it progresses. It is characterised by arthralgia and stiffness of the fingers, hands and wrists, but the development of synovitis is possible. The presentation can be difficult to distinguish from inflammatory arthritis or another systemic rheumatic disease. Subsequently, patients can develop a mildly inflammatory polyarthritis, which may be erosive[5]. Patients with SSc frequently have muscle involvement, ranging from disuse atrophy to inflammatory myositis, with elevation of creatine kinase concentrations. Joint effusion is rare, and analysis shows a fluid that is not very inflammatory. Inflammatory syndrome is not common – unlike in our case – and autoantibody status, despite possible rheumatoid factor positivity, is not associated with osteoarticular clinical or radiological signs. Moreover, tendon involvement is frequent and characteristic of the diffuse cutaneous form.

We found very few articles reporting on the coexistence of SSc and PMR, and the prevalence of these two disorders occurring together is not known. Orzano et al. reported four cases of SSc and PMR. In their report, the mean age at onset for SSc was 59 years and the mean age at onset of PMR was 64 years. Three out of four patients had the diffuse cutaneous form of the disease (dcSSc)[1] in contrast to our patient who presented a limited cutaneous disease; age at onset of SSc 46 years and age at onset of PMR 50 years.

The clinical picture of our patient was atypical since it was characterised by an asymmetry in the scapular girdle involvement, delayed onset of stiffness and inflammatory syndrome, and non-specific radiological findings that delayed management and motivated the performance of an ^{18}F-FDG-PET scan when the diagnosis of PMR was based on clinical findings. This examination also allowed us to rule out the presence of neoplasia or large-vessel vasculitis.

The coexistence of limited cutaneous SSc and giant cell arteritis has also been described[6]. Besides, PMR and giant cell arteritis are often concurrent and overlapping conditions[3]. In some patients, PMR is the presenting manifestation of giant cell arteritis, particularly in patients with predominant extracranial large-vessel involvement, which was not the case in the patient presented. Patients need to be carefully followed up as corticosteroids for PMR have been implicated in precipitating scleroderma renal crises.

CONCLUSION

The development of PMR during scleroderma is rare and the diagnosis challenging considering. ^{18}F-FDG-PET is a useful tool in diagnosis of PMR and its differential diagnosis.

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