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ADULT-ONSET STILL'S DISEASE PRESENTING WITH PROGRESSIVE MOTOR NEUROPATHY

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ABSTRACT

Introduction: Adult-onset Still's disease (AOSD) is a rare systemic autoinflammatory disorder characterized by dysregulation of the innate immune system and excessive cytokine production. Its presentation is often non-specific and heterogeneous, making diagnosis challenging and one of exclusion.

Case description: We report the case of an 18-year-old woman admitted with quotidian fever, frequent episodes of tonsillitis, intermittent skin rash and progressive distal lower limb weakness. Neurological assessment demonstrated progressive motor neuropathy. Laboratory findings revealed neutrophilic leucocytosis, significantly elevated inflammatory markers and hyperferritinaemia. Extensive investigation excluded infectious, malignant and autoimmune causes, and the patient met the Yamaguchi classification criteria for AOSD. Treatment with systemic corticosteroids, colchicine and the interleukin-1 receptor antagonist resulted in rapid and sustained clinical improvement.

Conclusion: This case highlights the diagnostic complexity of AOSD and emphasizes the importance of considering this diagnosis in patients with persistent fever, hyperferritinaemia and rare neurological manifestations.

KEYWORDS

Adult-onset Still's disease, hyperferritinaemia, fever of unknown origin, polyneuropathy, interleukin-1 inhibitors

LEARNING POINTS

- Adult-onset Still's disease should be considered in patients with fever of unknow origin and hyperferritinaemia.
- Neurological involvement, including polyneuropathy, may occur as an atypical manifestation.
- Early cytokine-targeted therapy is associated with rapid clinical improvement and better outcomes.

INTRODUCTION

Adult-onset Still's disease (AOSD) is a rare inflammatory condition within the Still's disease spectrum and is currently considered to share pathophysiological and clinical features with systemic juvenile idiopathic arthritis[1,2].

The disease is driven by activation of the innate immune system and cytokine-mediated inflammation, particularly involving interleukin (IL)-1, IL-6 and IL-18[1].

Clinically, AOSD typically presents with quotidian spiking fever, a transient salmon-coloured rash, arthralgia

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or arthritis and sore throat, accompanied by marked inflammatory laboratory abnormalities.

After systematic exclusion of infection, malignancy and autoimmune disease, the diagnosis relies on validated clinical criteria, particularly the Yamaguchi criteria[3].

Neurological involvement is uncommon and may further obscure and delay diagnosis.

CASE DESCRIPTION

An 18-year-old woman with no relevant past medical history was admitted because of progressive distal weakness of the lower limbs, associated with reduced osteotendinous reflexes.

She reported a quotidian spiking fever reaching 39°C and lasting more than one week, with nocturnal and early-morning peaks, recurrent episodes of tonsillitis and an intermittent maculopapular rash *(Fig. 1, 2 and 3)*.

During hospitalisation, the patient developed arthralgias with an inflammatory pattern.

Physical examination revealed progressive distal motor weakness with hyporeflexia, without sensory deficits. No lymphadenopathy or hepatosplenomegaly was detected.

Laboratory evaluation demonstrated neutrophilic leucocytosis (13.8 x10^3 leucocytes/µl [4–10 ×10^3], 91.4% neutrophils), significantly elevated C-reactive protein (352.1 mg/l [≤15]) and erythrocyte sedimentation rate (80 mm [<15]), and marked hyperferritinaemia, with a peak serum ferritin level of 3,300 ng/ml [11–306.8]. Blood and urine cultures, as well as infectious serologies, were negative. Autoimmune screening, including antinuclear antibodies and rheumatoid factor, was also negative.

A computed tomography scan of the chest, abdomen and pelvis revealed no evidence of infection, malignancy, lymphadenopathy or organomegaly. Transthoracic echocardiography excluded endocarditis and structural cardiac disease. Cerebrospinal fluid analysis showed normal findings.

Electroneuromyography demonstrated an axonal polyneuropathy with motor predominance, without sensory or autonomic involvement.

After exclusion of alternative diagnoses, the patient fulfilled the Yamaguchi classification criteria for AOSD, meeting four major (fever, arthralgia, typical rash and leucocytosis with neutrophilia) and two minor criteria (sore throat and negative tests for antinuclear antibody and rheumatoid factor)[3].

Treatment was initiated with oral prednisolone at a dose of 1 mg/kg/day, in combination with colchicine 1 mg daily. Due to systemic involvement and neurological manifestations, early therapy with the interleukin-1 receptor antagonist anakinra (100 mg daily) was introduced, in accordance with current recommendations for systemic Still's disease[2].

The patient showed rapid clinical improvement, with resolution of fever and arthralgias, and progressive recovery of motor function. Inflammatory markers, including ferritin levels, normalised during follow-up.



Figure 1. *Facial and cervical erythematous rash with, observed during active systemic inflammation.*



Figure 2. *Evanescent salmon-coloured maculopapular rash involving the lower limb, with irregular erythematous patches and areas of relative central sparing, observed during active systemic inflammation.*

DISCUSSION

Growing evidence supports the concept that AOSD and systemic juvenile idiopathic arthritis represent a single disease spectrum[1,2].



Figure 3. Palmar erythema with patchy erythematous macules involving the palms and fingers, observed during a febrile episode, consistent with inflammatory cutaneous involvement.

Hyperferritinaemia remains a useful marker of disease activity, although it is not included in formal classification criteria[4]. Emerging biomarkers, such as IL-18 and S100 proteins, may further improve diagnostic accuracy.

Neurological manifestations, including peripheral nervous system involvement, are rare in AOSD and have been described mainly in isolated case reports[4]. Their presence may delay diagnosis and complicate clinical management.

Early initiation of IL-1 blockade has demonstrated high efficacy in systemic and refractory disease, allowing corticosteroid sparing and improved clinical outcome[2].

REFERENCES

1. Feist E, Mitrovic S, Fautrel B. Mechanisms, biomarkers and targets for adult-onset Still's disease. *Nat Rev Rheumatol* 2018;**14**:603–618. doi: 10.1038/s41584-018-0081-x
2. Fautrel B, Mitrovic S, De Matteis A, Bindoli S, Anton J, Belot A, et al. EULAR/PReS recommendations for the diagnosis and management of systemic juvenile idiopathic arthritis and adult-onset Still's disease. *Ann Rheum Dis* 2024;**83**:1614–1627. doi: 10.1136/ard-2024-225851
3. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, et al. Preliminary criteria for classification of adult Still's disease. *J Rheumatol* 1992;**19**:424-430.
4. Gerfaud-Valentin M, Jamilloux Y, Iwaz J, Sève P. Adult-onset Still's disease. *Autoimmun Rev* 2014;**13**:708–722. doi: 10.1016/j.autrev.2014.01.058