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ADULT ONSET STILLS DISEASE WITH POSITIVE ANTINUCLEAR ANTIBODIES-A CASE REPORT

Internal Medicine

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ABSTRACT

Adult-onset Still's disease (AOSD) is a rare and complex systemic inflammatory disorder that presents with a myriad of symptoms, making diagnosis challenging. Here is a case report of a 43-year-old female presented with fever, joint pains, and rash. The patient exhibited a unique combination of positive antinuclear antibodies (ANA) and markedly elevated ferritin levels. Hence the patient was diagnosed with adult onset Stills disease based on Yamaguchi criteria. The patient was treated with corticosteroids and responded well to the treatment.

KEYWORDS

Adult onset stills disease, Arthralgias, Fever, Rash.

CASE REPORT

A 43-year-old female presented to our hospital with a history of high-grade fever of more than 39 degrees for 2 weeks which was associated with a sore throat, arthralgia involving wrists, shoulders, knees, ankles, and small joints of her hands, and a rash that appeared during fever spikes. On examination, there was a salmon-colored rash over the trunk and upper limbs (Figure 1). The patient's chest, cardiovascular, abdominal, and neurological evaluation was unremarkable.

Laboratory investigations showed a hemoglobin level of 10.6 g/L, markedly elevated C-reactive protein (CRP) at 148.10 mg/L, and an erythrocyte sedimentation rate (ESR) of 59 mm/hr. Liver function tests (LFTs) revealed transaminitis, with aspartate aminotransferase (AST) at 107 U/L and alanine aminotransferase (ALT) at 167 U/L. Procalcitonin levels were within the normal range (less than 0.5 ng/ml). Tests for viral infections, including IgM Epstein-Barr virus (EBV), cytomegalovirus (CMV), and Widal, were negative. Urine and blood cultures were sterile, and imaging studies (chest X-ray and abdominal ultrasound) were unremarkable.

Although antinuclear antibodies (ANA) were positive with a titer of 1:100, the patient did not have any manifestation of SLE, and other autoantibodies were negative. Notably, serum ferritin levels were markedly elevated at 10,007.10 ng/ml. The patient was diagnosed with adult onset stills disease as she fulfilled three major and two minor criteria (Yamaguchi criteria) with markedly elevated ferritin levels.

The patient was initiated on prednisolone 50 mg daily, resulting in rapid improvement of joint pains and the resolution of fever within one day. After three days of hospitalization, she was discharged in stable condition. A follow-up plan was established, including the tapering of steroids and regular monitoring to assess disease activity and any potential complications.

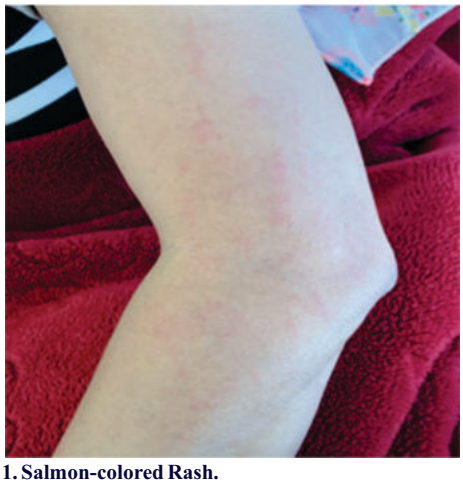

Figure 1. Salmon-colored Rash.

DISCUSSION

Adult-onset Still's disease (AOSD) is a remarkably rare condition with an estimated prevalence of 1.5 per 100,000 to 1,000,000 individuals, showing an equal distribution among genders (1). AOSD exhibits bimodal peaks of onset, typically affecting individuals between the ages of 15 to 25 years and between 36 and 46 years of age (2). Since its initial description by Bywaters in 1971, cases of AOSD have been gradually increasing in frequency (3). Despite ongoing research, the exact etiology of AOSD remains unknown, though some experts suggest that infectious agents, particularly viruses, may act as triggers for the disease in predisposed individuals (4).

Our patient exhibited a positive antinuclear antibody (ANA) without any other manifestations of systemic lupus erythematosus (SLE), and all other autoantibodies were notably negative. The presence of a positive ANA in AOSD is indeed rare but should not exclude the diagnosis of AOSD, highlighting the complexity of autoimmune disorders and the necessity for a comprehensive diagnostic approach (5,6).

Clinically, AOSD is characterized by a wide spectrum of symptoms. The most classic manifestations include daily fever (60%-100% of cases), a transient salmon-pink skin rash (60%-80%), sore throat (70%), and arthralgia (70%-100%), with fever and joint pain being the most prevalent symptoms. Other reported symptoms encompass myalgias (45%), lymph node enlargement (50%), splenomegaly (40%), hepatomegaly (30%), transaminitis (70%), hypoalbuminemia (76%), serositis with pleuritis (40%) or pericarditis (30%), abdominal pain (30%), pneumonitis (20%), and weight loss (27%) (7,8).

Management of AOSD is multifaceted, and various therapeutic strategies have been tried. In addition to symptomatic and supportive care, nonsteroidal anti-inflammatory drugs are used to alleviate fever, joint pain, and bone pain. Corticosteroids are essential in controlling systemic symptoms. Disease-modifying antirheumatic drugs (DMARDs) such as methotrexate, hydroxychloroquine, and azathioprine are considered depending on the disease's severity and the patient's safety profile. In our case, we opted for a treatment regimen involving nonsteroidal anti-inflammatory drugs, oral prednisolone, and supportive medications, which yielded a positive response and led to fever resolution.

For refractory cases, alternative treatments include the use of anti-TNF agents (infliximab, etanercept, and adalimumab), IL-1 inhibitors (anakinra, canakinumab, and rilonacept), and an IL-6 receptor antibody (tocilizumab), all of which have demonstrated efficacy in inducing remission in AOSD patients. Additional treatment modalities, such as plasma exchange and intravenous immunoglobulins, can be considered in patients with refractory AOSD (9).

CONCLUSION

This is a case of a 43-year-old female with AOSD highlights the diagnostic challenges of this rare systemic inflammatory disease. Early corticosteroid therapy can lead to clinical improvement and prevent

complications.

Abbreviations

AOSD-Adult onset stills disease, ANA-antinuclear antibodies, SLE-systemic lupus erythematosus, AST-aspartate transaminase, ALT-alanine transaminase, IGM-immunoglobulin M, EBV-ebstein barr virus, CMV-cytomegalovirus, LFT-liver function tests, CCP-cyclic citrullinated peptide, TNF-tumour necrosis factor, IL-interleukin.

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