Adult Onset Still's Disease

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INTRODUCTION

Adult Onset Still's Disease (AOSD) is an aetiologically unexplained systemic inflammatory disorder. AOSD occurrence is rare, prevalence being one in one lakh individuals.¹ The disorder mainly affects young adults mostly in the age group 15-45 years.² High spiking fever, fast vanishing rash, arthritis, raised leukocytes, high liver enzymes and hyperferritinaemia are the main characteristics of the disease. There are no specific tests for diagnosis. It is diagnosed after excluding other conditions and the differential diagnosis is usually long, which can include infections, neoplasm, autoimmune diseases, drug induced hypersensitivity reactions that can resemble AOSD's clinical presentation. There are many classification criteria,^{3,4} however Yamaguchi et al classification criteria are the most widely used.⁴ Non-steroidal anti-inflammatory drugs (NSAID), steroids, and immunosuppressive drugs are the usual treatment options. This report describes a female aged 24 who had pain in the throat, fever, joint pain, elevated liver enzymes, and hyperferritinaemia. After excluding other possible diagnoses, she was diagnosed with AOSD based on Yamaguchi criteria. The study demonstrated anaemia, elevation of inflammatory markers and hepatic enzymes, and negative serological and immunological results. Symptoms were overcome by the use of prednisolone.

PRESENTATION OF CASE

A twenty-four-year-old female reported with chief complaints of fever, throat pain since 2 months and joint pain since 2 weeks. Fever was high grade associated with chills, documented up to 103 F. The arthritis involved both small joints and large joints and morning stiffness was also present. Physical examination revealed throat congestion and cervical lymphadenopathy. Pallor and mild icterus. Abdominal examination showed palpable liver two fingers below right costal margin and mild splenomegaly. Other systemic examination was unremarkable.

On investigations haemoglobin (Hb) was 7.7 gm%, total leucocyte of 8.8 thousand/cumm, with polymorphs of 65%, platelet count of 340 thousand/cumm, malaria parasite by card test was negative, urine was normal, Liver function tests showed total bilirubin of 6.48 mg/dL, direct bilirubin of 4.2 mg/dL, SGOT of 275 u/l, SGPT of 581 u/l, alkaline phosphatase of 530 U/l. Kidney function tests, RA Factor, ASO titre, anti CCP antibody, antinuclear antibody, Hb electrophoresis were all normal. However, C-reactive protein (CRP) was positive, ESR-33 mm and serum ferritin was 1650 ng/ml. The serology testing for scrub typhus, leptospirosis, hepatitis A, B, C and E were negative. Lipid profile revealed total cholesterol 436 mg/dL, triglycerides 224 mg/dL, HDL cholesterol 28 mg/dL, LDL cholesterol of 363 mg/dL.

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An infectious disease possibility was considered and excluded; autoimmune markers were also negative. In view of the clinical features, raised inflammatory markers and transaminases with hyperferritinaemia AOSD was considered. Earlier the patient was treated with antibiotic empirically with no improvement. Since Yamaguchi criteria was fulfilled, the patient was given prednisolone 1 mg/kg/day, her symptoms improved, and she was discharged.

DISCUSSION

The pathogenesis of the AOSD disease is unclear; however, some studies suggest the involvement of genetic, infectious and environmental factors. TNF- α , interleukin IL-6 and IL-18 are raised, and are involved in AOSD pathogenesis. Fever, rash, sore throat and arthralgia/arthritis are usual clinical presentations. As in this patient, the fever usually exceeds 102 ° F. The usual rash is non-pruritic and occurs as salmon-pink, maculopapular outbreaks affecting the trunk and limbs, however it was absent in this patient. Though sore throat is a minor criteria, some study has given much importance to it, which may be present from the beginning of the illness, as was seen in this case. Other features of AOSD seen in this patient were hepatomegaly and splenomegaly. To diagnose AOSD 5 of the Yamaguchi criteria are to be present out of which at least two should be major criteria. The patient in discussion had 7 criteria, out of which 2 were major. Highly raised ferritin is characteristic of AOSD.⁵ Hyperferritinemia may be seen in up to 70% of patients,⁶ it is thought to be due to reticuloendothelial system or hepatic damage leading to cytokine secretion. Liver enzymes are elevated in about 75% of patients.⁷ Investigations ruled out all potential differential diagnosis for this case. The Yamaguchi criteria (1992) is the most agreed criteria having a sensitivity of 93.5 percent for diagnosis of AOSD.⁴

As an initial treatment in AOSD, NSAIDs and aspirin are prescribed, but were not used in this case in view of increased transaminases. In patients not responding to NSAIDs and having serositis, chronic anaemia or significantly elevated liver enzymes, prednisolone is an option.⁸ Some patients may require disease-modifying drugs such as methotrexate⁹ and biologics¹⁰ targeting TNF-alpha, IL1 and IL6. This patient was started on prednisolone and showed a good response.

CONCLUSIONS

A diagnosis of AOSD should be kept in mind, particularly in a patient with high-grade pyrexia of unknown origin, arthralgia/arthritis and skin rash, but only after excluding

differentials such as autoimmune disorders, infections, malignancy and vasculitis. High ferritin level may be a useful supportive marker in making AOSD diagnosis.¹¹ Management is challenging due to the complexity of the symptoms and the lack of accurate diagnostic markers.

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