CASE REPORT: A RARE CASE OF STILL DISEASE (AOSD)

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Abstract
Adult-onset still disease is an inflammatory disorder, characterized by quotidian (daily) fever, arthritis, and evanescent rash. It is a rare disease inflammatory disorder of unknown etiology.

Keywords: Fever, skin rash, polyarthritis, Yamaguchi criteria.

Introduction
Adult-onset Still disease (AOSD) is an uncommon clinical entity that predominantly affects predominantly females. One of the most common presentations of the disease is fever of unknown origin.

Early diagnosis can be difficult because fever of unknown origin is more commonly seen with many other conditions. Ambiguity in presentation and lack of serologic markers make diagnosis difficult.

AOSD is typically considered as a diagnosis of exclusion and a definitive diagnosis should be made by excluding infectious, malignant, and other connective tissue diseases.
Case Report

46 years old female name came to the Medicine OPD with complaints of: Fever associated with chills: since 4 months, Low back pain radiating to bilateral lower limbs, Joint tenderness since 1 month vomiting and rashes all over body since 4 days, cough and sore throat since a week

Past History: No other significant past history, No significant personal and family history on Examination – A febrile, BP: 110/70 mmHg, PR: 110 /m, RR: 20/m

Systemic Examination – CVS: Heart sounds normal, RS: bilateral inspiratory crep ts were present. P/A: soft, no tenderness noted, CNS: conscious, oriented to place time and time. Deep Tendon Reflexes - Knee jerk Absent, Bilaterally Ankle jerk Absent bilaterally Biceps, Triceps, Supinator present bilaterally planters Flexors bilaterally touch, pain, temperature and vibration normal Power 4+/5 in all four limbs Tone Normal in all four limbs.

Investigations – Haemoglobin : 9.7 gm /dl Total WBC : 19000 /cmm Platelet : 3,24,000


Discussion

AOSD was first described by Eric Bywaters in 1971.1 Pathogenesis of the disease remains unclear; however, observations suggesting the role of genetic, infectious and environmental factors have been published.2-4 There is a correlation between several cytokines in the pathogenesis of AOSD, including Tumor necrosis factor-alpha (TNF-α), interleukin (IL)-6 and IL-18. The levels of these cytokines are highly elevated in active AOSD
Patients with AOSD typically present with fever, rash, sore throat and arthralgia. The fever normally exceeds 39.0°C and highest temperatures are seen in late afternoon and early evening, as presented in this patient. The typical rash in AOSD is asymptomatic and is described as salmon-pink, maculopapular eruptions mainly affecting the trunk and extremities. Sore throat is one of the major signs of AOSD and may be associated with odynophagia. Arthralgia and arthritis mainly involving the knees, wrists, ankles and elbows have also been noted. The flare up of joint symptoms occurs during the febrile spikes. Carpal joints are the target of most destructive arthritis in AOSD.

The Yamaguchi criteria (1992), is the most widely used criteria to diagnose AOSD with a 93.5% sensitivity. In this criteria, there are 4 major and 4 minor criteria with 3 exclusion criteria. 1. Fever of at least 39°C for at least a week 2. Arthralgia or arthritis for at least 2-weeks. 3. Nonpruritic salmon colored rash on trunk/extremities 4. leukocytosis (10,000/microL or greater) Minor criteria 1. Sore throat, 2. Lymphadenopathy, 3. Hepatomegaly or splenomegaly 4. Abnormal liver function tests. 5. Negative tests for RF and ANA. Diagnosis requires at least 5 features, with at least 2 of these being major diagnostic criteria.

NSAIDs, corticosteroids, and DMARDs are the cornerstones of therapy for AOSD. NSAIDs were previously considered as the first-line medication. They have now been replaced by corticosteroids. Relatively high doses of steroids (equivalent to 0.5 to 1 mg/kg/d of prednisone) are required to induce clinical remission. In patients with inadequate response to corticosteroids, methotrexate is the best choice to control disease activity. There are a few studies available which showed limited success with anti-TNF drugs, interleukin blockade and intravenous immunoglobulin in AOSD.

References


