Adult Onset Still’s Disease Masquerading as Sepsis in an Asplenic Patient

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Abstract

Adult Onset Still’s disease (AOSD) is a rare inflammatory disorder of unknown etiology. It is a diagnosis of exclusion. We report a case of post-splenectomised man, presented with high grade fever, joint pain and body ache. Since Overwhelming Post Splenectomy Infection (OPSI) was the initial probable diagnosis, empirical antibiotic therapy was initiated. Evaluation to find a septic focus, autoimmune diseases ad malignancies was carried out which showed negative results. Since alternative diagnoses were excluded and the Yamaguchi’s criteria for AOSD was fulfilled, the patient was treated with IV steroids which resulted in rapid resolution of his symptoms. AOSD with asplenia is a unique condition because immunosuppressive therapy for AOSD may increase the risk of OPSI in such a patient. This is the first case report of AOSD in an asplenic patient from India.

Introduction

Adult onset Still’s disease (AOSD) is an auto-inflammatory disorder of unknown etiology with a prevalence of less than 1/100,000 people. It can manifest in multiple forms like pyrexia of unknown origin, serositis, aseptic meningitis, myocarditis, hemophagocytic syndrome and septic arthritis.¹ We report a case of an asplenic patient who presented with fever and polyarthralgia in whom AOSD was diagnosed. Immunosuppressive therapy was needed for the treatment of AOSD but the patient was on high risk of developing opportunistic infections because of his asplenic status. But the patient was treated successfully with intravenous steroids. This is the first case report of AOSD in an asplenic patient from India.

Case Report

A 39 year old man presented to our hospital with complaints of joint pain, body ache and intermittent high grade fever for one month and sore throat for four days. The joint pain involved both large and small joints. Despite analgesics there was no relief in pain. He also had swelling of the right knee and small joints of the hands. The patient had no other co-morbid illnesses. He had undergone splenectomy ten years ago following blunt injury to abdomen. Details of vaccination following splenectomy were not available.

On examination, the patient was conscious and oriented. He had a temperature of 101°F and blood pressure of 130/72 mmHg, with a regular pulse of 102/minute and respiratory rate of 15/minute. There was no rash or lymphadenopathy. Throat was found to be congested. Examination of abdomen revealed a left paramedian scar and no organomegaly. Locomotor examination showed tenderness over multiple joints (spine, shoulders, wrists, metacarpophalangeal joints, hip, knee and ankles). Swelling was noted in the small joints of hands and right knee. Rest of the systemic examination was normal.

Investigation reports at the time of admission were as follows: Hb-10.9 g/dl, TLC-24000/cu.mm, DLC-P90/L8/E2, Platelet count- 4 lakh/cu.mm, Urea-116 mg/dl, Creatinine- 2.1 mg/dl, Na-142 mmol/l, K- 5.2 mmol/l, Total bilirubin-2.8 mg/l , ALT-212 U/l, AST-198 U/l, ALP- 294 U/l, serum albumin-2.5 g/dl, serum globulin - 4g/dl. Creatinine kinase and lactate dehydrogenase were within normal limits.

Considering the patient’s asplenic status, Overwhelming Post Splenectomy Infection (OPSI) was the provisional diagnosis and empirical antibiotic therapy was initiated. Evaluation to find a septic focus or autoimmune diseases was done. Blood and urine cultures were sterile. Serological tests for dengue, chikungunya, EBV, hepatitis A-E, malaria, brucella, HIV and syphilis infections were negative. Tuberculin sensitivity test failed to show any significant induration. Bone marrow aspiration showed normoblastic erythropoiesis and myeloid hyperplasia. Rheumatoid factor and anti-nuclear antibodies were negative. Serum ferritin levels were 3230 ng/dl (normal: 40-200 ng/dl). Serum triglyceride and fibrinogen levels were normal. C-reactive protein (CRP) was raised (32 mg/dl). Erythrocyte sedimentation rate (ESR) was 95 mm at 1 hr. Abdominal ultrasonogram revealed no positive findings except asplenia. Radiographs of the chest, pelvis, hands and feet revealed no abnormality. No vegetations were seen on an echocardiogram.

There was no improvement in the symptoms after a week of treatment. Subsequent investigations showed persistently elevated total leukocyte count with neutrophilic predominance. Since the initial differential diagnoses were excluded, we looked for an alternate diagnosis. As the Yamaguchi’s criteria for Adult Onset Still’s Disease were fulfilled, it was decided to treat the patient with steroids. Due to the risk of OPSI, he was vaccinated against pneumococci, meningococci and hemophilus influenza b prior to treatment.

The patient was given three daily pulses of 500mg of intravenous methylprednisolone which resulted in rapid resolution of his symptoms. After a week of steroid therapy, he was discharged on 1 mg/kg of oral prednisolone and 15 mg/week of methotrexate. Investigations at the time

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inflammatory pathway. It has bimodal disorder caused by an aberrant innate.

Yamaguchi criteria (Table 1) have been devised. However, the least seven sets of diagnostic criteria could not be identified which might be negative RF/ANA. Typical Still’s rash led to the diagnosis of AOSD. He was treated with methylprednisolone and anakinra following which he showed marked improvement.7

In healthy individuals, 50-80% of ferritin is glycosylated. Several studies have shown that the level of glycosylated ferritin is low in AOSD patients (<20%). The reason behind decreased glycosylated ferritin could be the saturation of glycosylation process due to hyperferritinemia or decreased clearance of non-glycosylated proteins by histiocyte-macrophage system.8 But as this test is not readily available, we could not get this test done in our patient.

Jaqua et al reported a similar case of AOSD in a 26-year-old soldier who presented with fever and arthralgia. He had undergone splenectomy for ITP. Initially he was treated with empirical antibiotics but later developed a typical Still’s rash which led to the diagnosis of AOSD. He was treated with methylprednisolone and anakinra following which he showed marked improvement.7

NSAIDs, corticosteroids, and DMARDs are the cornerstones of therapy for AOSD. NSAIDs were previously considered as the first-line medication.8 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.9 There are a few studies available which showed limited success with anti-TNF drugs, interleukin blockade and intravenous immunoglobulin in AOSD.10

Our patient showed remission with steroids. In an asplenic patient, long-term corticosteroids are relatively contraindicated, given the potential predisposition to infection. So, he was discharged on tapering doses of oral prednisolone. Methotrexate was started after confirming normal liver and kidney functions. In future, this patient’s fever could be indicative of OPSI or flare-up of AOSD. So he was advised to obtain medical care immediately.

Conclusion

The presentation of fever with asplenia made us focus on OPSI by an encapsulated organism. But when the patient did not improve with initial management, it made us look for alternative diagnoses thus leading to the rare diagnosis of AOSD. Keeping an open mind while evaluating the patient can prevent an unusual disease from being misdiagnosed.

References