A CASE OF ADULT ONSET STILL’S DISEASE: A REVIEW ON DIAGNOSTIC WORKUP AND TREATMENT MODALITIES

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ABSTRACT Adult Onset Still’s Disease (AOSD) is a rare systemic inflammatory idiopathic disease that presents as Fever of Unknown Origin (FUO) (in 5-10% patients) accompanied by systemic manifestations. We report an interesting case of a 38-year-old Male Indian resident who presented with a fever of unknown origin (FUO) along with skin rash, sore throat, and arthralgia accompanied by systemic manifestations. After extensive workup, we ruled out the potential differential diagnosis, and a diagnosis of AOSD was suspected due to his clinical presentation and fulfilment of Yamaguchi and Fautrel criteria. This case report discusses the case history, clinical manifestations, diagnostic workups, and differential diagnoses.

KEYWORDS Adult Onset Stills Disease, AOSD

Introduction

Adult Still’s Disease is also known as AOSD or Wissler-Fanconi Syndrome. It is a chronic inflammatory disorder characterized by high fever, joint pain, and nonpruritic rash. Still’s a disease was initially described by George F. Still in 1896 as a form of chronic joint disease in children, resembling rheumatoid arthritis in adults[1]. It is a rare inflammatory disorder that affects the entire body (systemic disease). It is a chronic, potentially disabling condition. It can also be defined as the adult form of systemic juvenile Rheumatoid Arthritis. In 1971, EG Bywaters described the first series of 14 adults with the same symptoms as those seen in pediatric Still’s disease, defining thus the adult-onset Still’s a disease (AOSD)[2]. The aetiology of AOSD remains unclear, and viral infections, genetic factors, and immune dysregulation, including cytokine-mediated inflammation, and dysregulated apoptosis, have all been implicated in the development of this disease.[3-7] The characteristic features include the triad of Fever (more than 102.2°F, which spikes in the late afternoon or early evening), Salmon coloured bumpy rashes and Arthralgia. The salmon-pink rash usually develops during the fever spike and disappears quickly. Additional findings include sore throat, splenomegaly, hepatomegaly, lymphadenopathy, pericarditis or myocarditis, pleural effusion, heart and lung involvement, which can cause difficulty breathing and chest pain. [8-11] Diagnosis of AOSD is based on Yamaguchi or Fautrel criteria[12] which is as follows: Diagnosis is further confirmed after excluding the malignancy, other rheumatic diseases, and infectious causes and if the patient completes ≥ 5 criteria which must include 2 major criteria.

Case report

A 40-year-old Indian male presented to the casualty department with a 6-month history of high-grade fever, sore throat, joint pain with stiffness, dry cough, and limitation of daily activities. His fever was accompanied by a non-pruritic macular, diffuse, erythematous blanching skin rash on his trunk, abdomen, back, and palms which tends to disappear with subsiding fever, myalgia, arthralgia of bilateral ankles, wrist and knees, profuse sweating at night and loss of weight since last 6 months. He denies having joint stiffness in the mornings, blurry vision, eye pain, oral ulcers, headache, recent travel, burning micturition, and sick contacts. He had no significant past medical history, was not on any medication previously, and had no significant family

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Major Criteria | Minor Criteria
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Fever ≥ 39°C lasting ≥ 2 weeks | Sore throat
Arthralgia or arthritis lasting ≥ 2 weeks | Lymphadenopathy
Typical non-pruritic salmon-pink rashes | Splenomegaly
Leukocytosis ≥ 10,000/mm³ with granulocytes ≥ 80% | Abnormal Liver Function Tests

Table 1: Initial blood investigation and results.

<table>
<thead>
<tr>
<th>Tests CBC</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>10.3 g/dl</td>
</tr>
<tr>
<td>Total Leucocyte count</td>
<td>18,200 /cmm</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>92%</td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>5%</td>
</tr>
<tr>
<td>Eosinophils</td>
<td>1%</td>
</tr>
<tr>
<td>Platelets</td>
<td>2,98,000</td>
</tr>
<tr>
<td>ESR</td>
<td>45 mm/hr</td>
</tr>
<tr>
<td>Serum Ferritin</td>
<td>7117.92 ng/ml</td>
</tr>
</tbody>
</table>

and personal history. He had no known allergies. On physical examination, the patient was febrile with a temperature of 39°C, tachycardia with a heart rate of 120 beats per minute, respiratory rate of 20 breaths per minute, and blood pressure of 120/70 mm Hg. He also had cervical, axillary, and splenic lymphadenopathy and an inflamed throat but without any exudates. The musculoskeletal system showed minimal tenderness in bilateral ankles, wrist, and knees with normal active and passive range of motion. There were no signs of active synovitis in any of his joints. Arthralgia is more severe in the knee than in the wrist and ankle. On examination of the abdomen, splenomegaly and hepatomegaly were seen. Cardiovascular, respiratory, and neurological examination was insignificant. He was admitted to the hospital, and his initial workup revealed elevated acute phase reactants (Erythrocyte Sedimentation Rate (ESR): 45 mm/hr, C-reactive protein (CRP): 78.4 mg/l and Serum Ferritin: Significantly raised-7117.92 ng/mL), mildly elevated liver function tests, normocytic anaemia with slightly increased white cell count (18,200 cells/cu.mm) with neutrophilic predominance (92%), and normal platelet count. Rapid strep throat test, hepatitis, HIV, urine analysis, blood, and throat cultures were all negative. Rheumatoid factor (RF), Anti Nuclear Antibody (ANA), Brucella antibodies, IgG, and cyclic citrullinated peptide (Anti-CCP) antibody were negative too. Chest X-Ray showed no acute infiltrates. Laryngoscopy and Nasal endoscopy did not show any infective focus. The patient received a course of antibiotics (Ceftriaxone) during hospitalization, which did not resolve his symptoms. An abdomen and pelvis CT was done, which was normal except for mild splenomegaly. An extensive ID workup was ordered, including fungal serology and gallium scan, all of which were normal. Haematology was consulted for the patient’s FUO and normocytic anaemia. Haematological conditions such as leukaemias, lymphomas, and hemophagocytic lymphohistiocytosis were ruled out due to the absence of physical and laboratory findings. A 2D echo, ordered to rule out endocarditis, came back as a normal study. Several investigations, including serum protein electrophoresis and urine protein electrophoresis, were nondiagnostic for the patient’s FUO and anaemia. A bone marrow biopsy with flow cytometry was finally ordered. The bone marrow study showed a normocellular pattern with adequate storage of iron and no evidence of fibrosis or lymphoproliferative disorder. Rheumatology was consulted for his FUO in relation to arthralgia and myalgia. After ruling out sickle cell anaemia, tuberculosis, and other infectious causes, a diagnosis of adult-onset Still’s disease was made based on the Yamaguchi or Fautrel’s criteria.[12]

He was started on oral prednisolone at 1 mg/kg and antipyretics and analgesics were started as supportive care. He was discharged on oral prednisolone 40 mg per day and tramadol. The patient came for follow-up after 1 month; the patient was symptomatically better with no episodes of fever or joint pain, and rash. So prednisolone was tapered, and he was started on 15 mg weekly methotrexate. During follow-up, the patient was on 5 mg prednisolone per day, and there was no episode of fever or arthralgia. CBC was repeated for 2 months, and it showed reduced ESR and leukocytosis. Therefore the patient was continued on 5 mg prednisolone per day and 7.5 mg methotrexate per week. The patient is regularly under follow-up to date.

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**Conflict of interest**
There are no conflicts of interest to declare by any of the authors of this study.
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References