INTRODUCTION

A 19 year old female presented to our hospital with fever, rashes over chest, joint pains, body aches and throat pain for the last 15 days. Fever was high grade; persistent. Rashes were over the chest and non-pruritic. The joint pain involved both large and small joints. On examination, patient was febrile having fever of 101°F, pulse rate of 94 beats/min and respiratory rate of 23 breaths/min. Evanescent nonpruritic macular rash was seen over chest. There was generalised lymphadenopathy involving cervical, axillary and inguinal lymph nodes. Investigations revealed CBC: Hb:9.2g/dl, TLC: 19,200/mcl, platelets: 1,85,000/mcl, ESR w 9925ng/ml, folic acid: 7.09ng/ml, vitamin B12: 244.07pg/ml, LDH: 787IU/L. CXR was normal, HBsAg, anti HCV, anti HIV: negative, Whole body PET scan revealed FDG avid b/l cervical, axillary, abdominal, pelvic and inguinofemoral lymphadenopathy with hepatosplenomegaly (19cm liver, spleen mildly enlarged), mild diffuse uptake in spleen and bone marrow. Bone marrow biopsy was done to rule out underlying malignancy, it showed macrophages and increase in myeloid series. Bone marrow aspiration showed cellular marrow, myeloid preponderance, 4% erythroid cells and 5.2% large lymphoid cells and haemophagocytosis. Bone marrow culture was sterile.1 cervical and 2 left axillary LNs were excised and sent for HPE which was negative for malignancy.ANA, ANA profile, RF, Anti CCP: negative. Patient was started on steroid: Inj. Solumedrol 40mg i/v BD following which her fever subsided, joint aches got relieved.

DISCUSSION

Still’s disease is named after doctor George Still who described it in children. AOSD was initially described by Bywaters in 1971 as a distinct clinical entity in adults. The exact pathogenesis is unknown. The important step is IL18 mediated macrophage and neutrophil activation as evidenced by upregulation of CD64 in these patients. Serum levels of TNF alpha, INF gamma, IL 1, 6, 8 and 18, SIL-2R are also elevated in active disease. The classic manifestations are fever with rash (transient, nonpruritic, salmon coloured, macular or maculopapular often observed during fever spikes commonly

Case Report

ADULT ONSET STILL’S DISEASE

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ABSTRACT

Adult onset Still’s disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology characterised by persistent high grade fever, arthritis and an evanescent (salmon colored) rash. Diagnosing AOSD is difficult due to nonspecific symptoms and absence of characteristic serological markers. Our case is of an 19 yr old female with no known comorbidities and no underlying infection, malignancy or other connective tissue disease meeting the diagnostic criteria (Yamaguchi criteria) for AOSD. AOSD is a diagnostic dilemma for physicians as it presents with non-specific symptoms and an increased awareness is warranted among physicians.

Key Words:

Adult onset Still’s disease, AOSD, Yamaguchi criteria

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on trunk and proximal extremities), sore throat and arthralgia (involving knees, ankles, wrists and elbows). Common laboratory abnormalities are elevated ESR, neutrophilic leucocytosis, thrombocytopenia, raised serum ferritin (>2000ng/ml). Glycosylated ferritin is typically decreased in these patients. Other less common lab findings include serum albumin < 3.5mg/dl, elevated AST/ALT and anaemia of chronic disease. RF and ANA are usually negative. Radiographic results are generally normal in the early phase and may be helpful in the late and chronic phase with worsening erosions and joint space narrowing (carpometacarpal joints are more commonly involved than tarsometatarsal joints). Yamaguchi criteria is most widely followed as it is 96% sensitive and 92% specific.

**Table 1 - Classification criteria for adult-onset Still’s disease proposed by Yamaguchi et al**

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
</tr>
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<tbody>
<tr>
<td>Temperature of &gt; 39°C for &gt; 1 wk</td>
<td>Sore throat</td>
</tr>
<tr>
<td>Leukocytosis &gt; 10 000/mm³</td>
<td>Lymph node enlargement</td>
</tr>
<tr>
<td>Typical rash</td>
<td>Splenomegaly</td>
</tr>
<tr>
<td>Arthralgia &gt; 2 wk</td>
<td>High transaminases</td>
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<tr>
<td></td>
<td>Negative ANA, RF</td>
</tr>
</tbody>
</table>

After excluding infections, malignancies, and other rheumatic diseases, adult Still’s should be considered if 5 criteria (2 of which being major ones) are met. ANA = antinuclear antibody; RF = rheumatoid factor

Our patient fulfilled 4 major and 4 minor criteria. Another criteria was proposed by Fautrel et al in 2002 which contained glycosylated ferritin as a new marker (20% or less: diagnostic). The disease pattern is divided into 3 distinct types: monocyclic/ self-limiting, polyyclic/ intermittent (2 or more episodes separated by symptom free remission period lasting for a minimum of 2 months), chronic articular pattern characteristic by severe articular manifestations causing joint destruction. The major diseases that should be excluded include infections, granulomatous diseases, malignancy and connective tissue diseases. First line treatment is corticosteroids. DMARDs (MTX, cyclophosphamide, azathioprine and cyclosporine) are often used for maintenance therapy of the disease. Cyclosporine can be used for AOSD patients presenting with macrophage activation syndrome (MAS) as seen in our case. Among the 3 IL 1 antagonists: Anakinra, Canakinumab and Rilonacept, Anakinra has been used more frequently. IL1 inhibition is considered the mainstream of treatment for refractory AOSD.

**CONCLUSION**

Adult onset Still’s disease can be missed due to nonspecific symptoms and lack of a characteristic diagnostic marker. It is a diagnosis of exclusion, final diagnosis is made after ruling out viral infections like HIV, hepatitis B and C, EBV, CMV, Coxsackie, rubella, Tb, malignancy (leukemias, lymphomas), granulomatous diseases like Sarcoidosis, Crohn’s disease and connective tissue diseases: SLE, MCTD, PAN, Wegener’s granulomatosis, Takayasu arthritis. It is an important cause of PUO (pyrexia of unknown origin) and needs to be diagnosed and treated timely for better outcomes.

**References**


