

# Juvenile Onset Still's Disease Presenting as Pyrexia of Unknown Origin: A Case Report

<sup>#1</sup>Himanshu Juneja, <sup>#2</sup>Surekha Dabla

<sup>#1</sup> Sr. Resident, PGIMS, Rohtak, Haryana, India

<sup>#2</sup> Professor, PGIMS, Rohtak, Haryana, India

## Abstract

*Juvenile onset still's disease is a rare clinical entity with unknown etiology characterized by fever, evanescent rash, arthralgias and other systemic presentations. This report described a 15 year old male who presented with high grade fever, sore throat, rash, arthralgias, raised liver enzymes and hyperferritinemia. He was diagnosed to have Still's Disease based on Yamuguchi criteria after the exclusion of other potential diagnoses. The patient started on methotrexate, etoricoxib and prednisolone and responded well to treatment.*

**Keywords** --- Still's disease, Yamuguchi criteria, Systemic onset Juvenile Idiopathic Arthritis.

## I. INTRODUCTION

Juvenile onset still's disease is a rare inflammatory disorder of unknown etiology usually present as pyrexia of unknown origin characterized by high spiking fever, salmon colored rash, joint pain, and multiorgan involvement<sup>1</sup>. The diagnosis is one of exclusion. Here we report a case of 15 year old male who presented with fever of unknown origin.

## II. CASE PRESENTATION

A 15 year male not a known case of any chronic illness presented with fever high grade, intermittent, associated with chills and rigors, sore throat, generalized bodyache and diffuse arthralgias since 1 week. This was preceded by red pruritic maculopapular rash mainly over the trunk and extremities. Both medical and family histories were unremarkable. On admission patient was conscious, oriented to time/place/person. His vitals were Temp. 102 °F, pulse rate was 110/min, blood pressure was 130/70 in right arm supine position and with a respiratory rate of 25 breaths/min. On physical examination salmon colored maculopapular rashes were present on trunk and all four extremities (figure 1 & 2). Cervical and axillary lymph nodes were palpable. There was mild congestion on the posterior pharyngeal wall. Per abdomen revealed just palpable liver (liver span was 15cm), no splenomegaly. No other significant finding was present on systemic examination.

Laboratory findings revealed hemoglobin 12.3 g/dL, WBC count 11500/mm<sup>3</sup> (neutrophils 88%, lymphocytes 10%, monocytes 1%, eosinophils 1%), platelet count 2.0 lac/mm<sup>3</sup> and erythrocyte sedimentation rate 80mm/h, C-reactive protein 25.0

mg/L, ASO titer were <100 U/L. Liver enzymes were elevated ( aspartate transaminase 486 U/L, alanine transaminase 179 U/L, alkaline phosphatase 217 U/L). Renal and coagulation profiles were normal. Urine routine examination revealed traces of proteins and few epithelial cells, with no RBC/WBC cast were seen on microscopic examination. On chest x-ray no abnormality was seen. Empiric treatment with antibiotics (ceftriaxone and amikacin) and antipyretics were commenced immediately. Patient did not respond to the treatment and continued to have spiking fever for next 1 week. Further laboratory investigations revealed elevated serum ferritin levels (>1000 ng/ml). Rheumatoid factor (RF) and antinuclear antibody (ANA) were negative. Serological tests for Dengue, Malarial parasite, Salmonella typhi and paratyphi, Ebstein-barr virus, Scrub typhus, Brucella were negative. Viral markers for HIV, HbsAg, and HCV were negative. Tubercular skin testing was negative. No growth of aerobic and/or anaerobic bacteria was seen on blood culture. CECT thorax and abdomen revealed mild pericardial effusion, hepatomegaly with periaortic lymphadenopathy. Bone marrow aspiration revealed non-specific myeloid response.

Based on his clinical features and laboratory investigations, he was diagnosed to have Still's Disease using the Yamaguchi's criteria. He was started on methotrexate 5 mg once a week, etoricoxib 120 mg/bd and prednisolone 40 mg/od. Over the next few days, the patient became afebrile, arthralgia improved and he was able to ambulate himself without assistance. The patient was discharged on prednisolone 30 mg daily and etoricoxib 120 mg once daily. During follow up; patient was afebrile and steroids were tapered off.

## III. DISCUSSION

Juvenile onset still's disease also known as Systemic onset Juvenile Idiopathic Arthritis (JIA) is similar to adult onset still's disease. By definition juvenile form occurs before the age of 16 years. Still's disease is an inflammatory multisystemic disease of unknown etiology. Although exact etiopathogenesis of Still's disease is not known, various host factors like HLA alleles like HLA-DR2, DR4 and environmental factors like infections have been supposed to play some role in etiopathogenesis of disease. An underlying severe autoinflammatory response is the hallmark of still's disease<sup>3</sup>. Its main clinical features are combination of high grade fever,

pink-salmon colored cutaneous rash appearing on trunk and extremities and joint pain (Still's triad). Other clinical manifestations like hepatosplenomegaly, lymphadenopathy, serositis and aseptic meningitis can occur. Severe disease complications like pericarditis, endocarditis, hemolytic anaemia and macrophage activation syndrome (MAS) can occur. Important laboratory findings are leukocytosis with predominance of neutrophils, negative testing for rheumatoid factor (RF), and antinuclear antibody (ANA), high serum

ferritin levels as well as elevated liver enzyme levels<sup>2</sup>. The exclusion of other potential diagnosis is one of key steps when the diagnosis of Still's disease is considered. The Yamaguchi criteria is the most widely used criteria to diagnose still's disease. In these criteria, there are 4 major and 4 minor criteria with 3 exclusion criteria as shown in the table 1. Five or more criteria must be met in order to make a diagnosis of Still's disease, including 2 or more major criteria, after excluding infections, malignancies and autoimmune disease.



Figure 1& 2:- Salmon Coloured Maculopapular Rash on Trunk and Limb

Table 1:- Yamaguchi's Classification Criteria<sup>4</sup>

Major Criteria	Minor Criteria	Exclusion criteria
<ul style="list-style-type: none"> <li>▪ Fever &gt;39 °C intermittent for &gt;1 week</li> </ul>	<ul style="list-style-type: none"> <li>▪ Sore throat</li> </ul>	<ul style="list-style-type: none"> <li>▪ Infections</li> </ul>
<ul style="list-style-type: none"> <li>▪ Arthralgias &gt;2 weeks</li> </ul>	<ul style="list-style-type: none"> <li>▪ Lymphadenopathy and or splenomegaly</li> </ul>	<ul style="list-style-type: none"> <li>▪ Malignancies</li> </ul>
<ul style="list-style-type: none"> <li>▪ Typical rash</li> </ul>	<ul style="list-style-type: none"> <li>▪ Abnormal liver function</li> </ul>	<ul style="list-style-type: none"> <li>▪ Inflammatory disease</li> </ul>
<ul style="list-style-type: none"> <li>▪ WBC &gt;10000 (PMN &gt;80%)</li> </ul>	<ul style="list-style-type: none"> <li>▪ RA factor (Negative)</li> <li>▪ ANA (Negative)</li> </ul>	

As far treatment is concerned nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids are first line treatment for still's disease. Disease modifying antirheumatic drugs (DMARDs) such as methotrexate, azathioprine, cyclosporine are considered in steroid non-responsive cases<sup>5</sup>.

### REFERENCES

- [1] Ramanan AV, Grom AA. Does systemic-onset juvenile idiopathic arthritis belong under juvenile idiopathic arthritis. *Rheumatol* 2005;44:1350-3.
- [2] Luthi F, Zufferey P, Hofer MF. Adolescent-onset Still's disease: characteristics and outcome in comparison with adult-onset Still's disease. *Clin. Exp. Rheumatol.* 2002; 20: 427–30.
- [3] Hinks, A; Cobb, J; Prahalad S; Sudman, Marc; Glass, David; Langefeld, Carl; Thomson, Wendy; Thompson, Susan et al. Dense genotyping of immune loci using Immuno Chip identifies 14 new susceptibility loci for juvenile idiopathic arthritis". *Nature Genetics* 2013; 45:664–9.
- [4] Yamaguchi M, Ohta A, Tsunematsu T. Preliminary criteria for classification of adult Still's disease. *J. Rheumatol.* 1992; 19:424–30.
- [5] De Witt, Morgan; Kimura, Yukiko; Beukelman, Timothy; Nigrovic, Peter A. Onel K et al. Consensus treatment plans for new-onset systemic juvenile idiopathic arthritis. *Arthritis Care & Research* 2012; 64:1001–10.