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Case Report

Adult Onset Still's Disease presenting as Pyrexia of Unknown Origin

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***ABSTRACT*:**
Adult onset Still's disease (AOSD) is a chronic multi-system inflammatory disorder characterized by high spiking fever, polyarthralgia and skin rash. Lymphadenopathy and hepatosplenomegaly is another prominent feature of adult onset Still's disease. We describe a 19 years old lady presented with fever, skin rashes and polyarthritis for 3 months. Examination revealed fever, typical skin rash, lymphadenopathy and polyarthritis. On investigation there was neutrophilic leukocytosis, high ESR, high ferritin level but RA test as well as ANA test were negative. History, clinical examinations and laboratory findings fulfill Yamaguchi criteria diagnostic of AOSD. With proper treatment, now she is free of symptoms and leaving a healthy life.
Key words: Adult onset Still's disease, Polyarthralgia, Salmon colored evanescent rash.

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INTRODUCTION:
Adult onset Still's disease is systemic onset juvenile idiopathic arthritis. It is a rare systemic auto immune disease. Its major clinical manifestations include high spiking fever, sore throat, musclepain, polyarthralgia, salmon colored evanescent rash, hepatosplenomegaly, lymphadenopathy and neutrophilic leukocytosis[1,2]. It is a chronic systemic inflammatory disorder of unknown etiology and is a diagnosis of exclusion. AOSD is rare and has a bimodal age distribution in all ethnic groups with peaks at 15-25 and 36-46 years of age in both sexes with an incidence of 0.16 cases /100000 persons /year[1,2] Recently we diagnosed a case of AOSD in our hospital, who became complete symptoms free with proper treatment, though it is non curable.

CASE REPORT:
A 19 year old female presented to our department with fever since 5 months duration. She started getting fever from 10/4/2017. Fever was of high grade, continuous type with no diurnal variation.
She developed rash 15 days later. It first appeared on trunk followed by upper and lower limbs. Rash was erythematous, bumpy, macular and pruritic in nature (Fig.1). She also complained of pain during swallowing for 8 days. Patient also complaints of joint pains in all major joints since 3 months. No morning stiffness. Two months later she also complains of vomiting, precipitated after taking food and vomitus contained food particles. For these complaints she was admitted and treated with antibiotics and brief course of glucocorticoids for 10 days in May 2017.
Fever subsided for one week and then recurred. She was transferred to a multi-speciality hospital and had further investigations but fever did not respond. No H/O headache, chest pain, breathlessness, pain in abdomen, burning micturition. H/o jaundice 5 year back that lasted approximately 2 weeks. No past history of congenital disorder, epilepsy, DM, HTN.

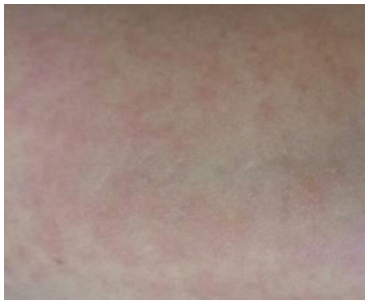

Figure 1: Salmon pink rash clearly visualized on forearm. The characteristic pinkish redness can be easily indentified. Rash was erythematous, bumpy, macular and pruritic in nature.

On examination, vitals stable, congestion of posterior wall pharyngeal wall seen, cervical lymphadenoapthy present. On per abdomen examination, splenomegaly is present. Otherwise, all his system examinations were within normal limits.
She had haemoglobin of 10.3g/dl, tlc of 18,900/micro-lit, with neutrophilia and platelets of 1,99,000/micro-lit. Her liver function tests were slightly deranged. To rule out infectious causes Dengue antigen test, Malaria antigen test, monospot test (Paul bunnel), Weilfelix, Chikengunya, Widal, HIV, HBs-Ag, Brucella antigen test done. All were negative. To rule out other inflammatory disorders RA factor, Anti- CCP, Procalcitonin done. All were negative. Serum ANA was negative.No blast abnormality on lymph node biopsy. Since no obvious infectitious, inflamatory or malignant cause for her condition was forthcoming we thought of AOSD. Serum ferritin was high. As she was fulfilling Yamaguachi criteria diagnosis of AOSD was established. (she is having 4 major and 3 minor features). She was started on prednisolone 40mg daily, after which she became afebrile for the first time.The patient showed considerable improvement and was discharged home after one week on prednisolone 40mg daily. She was advised to taper dose of prednisolone 5mg per week.on follow up 2 weeks later she is symptom free and living a healthy life.

DISCUSSION:
Adult Onset Still Disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology and pathogenesis that typically affects young adults aged between 16-35 years. It includes three variants of arthritis found in pediatric age group in the form of rheumatoid arthritis (RA), chronic fibrous rheumatism (Jaccoud's arthropathy) and systemic onset type (Still's disease) described by George Friderick Still (1897). It has been recognized with increasing frequency since the first description by Bywaters in 1971[3]. AOSD is a rare disease affecting all races. The demographic study revealed a higher prevalence of female patients and a relatively younger onset of
Exclusions:
I. Infections (especially, sepsis and infectious
II. Malignancies (especially, malignant lymphoma)
III. Rheumatic diseases (especially, Polyarteritis Nodosa and rheumatoid vasculitis with
Extraarticular features)

- Classification of AOSD requires 5 or more criteria including 2 or more major criteria.
- Any disease listed under "Exclusions" should be excluded.

Laboratory features of the disease are; Increased serum levels of CRP and ESR, leukocytosis, liver dysfunction, negative results for both rheumatoid factor and antinuclear antibodies and an increased ferritin levels[4]. Increased serum ferritin level is a nonspecific finding and should not be regarded as a diagnostic test. That ferritin may be helpful for monitoring disease activity during treatment[8]

male patients compared with female. The reported prevalence is one per 100.000 adults aged between 16 and 35 years[4]
Clinically, most of the patients with AOSD present with fever, sore throat, arthralgia, arthritis, and/or skin rash, but some patients may also have present with conditions such as lymphadenopathy, hepatosplenomegaly, and/or serositis. The fever typically higher than 39°C, starts suddenly and could present as FUO alone. Joint pain occurs in two thirds of patients. The arthralgia starts concomitantly with fever, involves any joint, and may migrate at the beginning and become more stable during the course of the disease. The skin rash consists of small discrete, nonpruiritic, salmon-pink macules or maculopapules, which are transient and mainly visible during fever.
AOSD is a diagnosis of exclusion. Differential diagnoses are infections, such as endocarditis and deep-seated occult infections, or neoplastic etiology, especially lymphomas and autoimmune diseases like vasculitis and polymyositis. These conditions should be excluded before AOSD is diagnosed.[5] There is strong association between cytokine and chronic articular disease in AOSD.
Tumor necrosis factor (TNF), interleukin (IL)-18 were increased in both types of AOSD, even in remission. Soluble receptors, IL-4, IL-18 level and IL-8 were correlated with disease activity[6]
The Yamaguchi criteria 1992 (specificity 92% and sensitivity 96%) is the most widely used criteria to diagnose AOSD[7].

Major criteria:	**Minor criteria:**
Fever of 39˚c or higher, lasting 1 week or longer	Sore throat
Arthralgia lasting 2 weeks or longer	Lymphadenopathy and/or splenomegaly
Atypical rash	Liver dysfunction
Leukocytosis (10,000/mm3or greater) including 80% more of granulocytes	Negative RF and negative ANA

The treatment of AOSD is mainly centered on the use of NSAIDs, steroids, and DMRDs[9]
NSAIDs and prednisone are used as first-line agents in the treatment. After the diagnosis is established, steroids are usually required for symptom control and response is often dramatic. Patients with visceral involvement may achieve a response with an intravenous infusion of high-dose methylprednisolone.[10,11] Methotrexate is effective in controlling disease activity and allows for steroid dose sparing in AOSD.[12] The potential efficacy of interleukin-I receptor antagonist (anakinra), TNF blockers, immunoglobulins, and cyclosporine has been reported.

CONCLUSION:
AOSD is a rare systemic inflammatory disorder of unknown aetiology and pathogenesis. It should be considered in the course of evaluating patients with triad of fever, rash and arthritis with non-invasive workup for FUO, along with documentation of fever pattern and

observation of patient over a minimum of at least 6-8 weeks. Any multisystemic disorder which is RAF or ANA negative should raise suspicion of AOSD.

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