Acta Scientific MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 7 Issue 1 January 2023

Case Report

Adult - Onset Still's Disease - Case Report

Tenzin Gyaltsen*, Ajay Gowda, Sarath Chandran Kr, Satyajit Hajong, Nitajan Nongtdu and Gido Pertin

PGTs in Department of General Medicine, Regional Institute of Medical Sciences, Imphal, India

***Corresponding Author:** Tenzin Gyaltsen, PGTs in Department of General Medicine, Regional Institute of Medical Sciences, Imphal, India.

DOI: 10.31080/ASMS.2022.07.1432

Received: November 29, 2022

Published: December 28, 2022

Abstract

Background: Adult onset still's disease (AOSD) is a rare multisystemic inflammatory disorder with an unknown etiology. Many theories have been put forward to understand the pathogenesis of the disease. The sign, symptoms and severity of the disease vary among different individuals. The affected person generally present with arthritis, spiking fever with pink colored rash, muscle, joint pain and with other systemic presentations. Diagnosis is generally made after thorough clinical examination which requires exclusion of other conditions which mimics same presentation such as infection, malignancy and autoinflammatory condition before labeling a patient to be affected with AOSD.

With the above background, we report a case of 42 yrs. female who presented with joint pain, fever, sore throat and pink colour rash predominantly on trunk and lower limbs which generally decreased whenever fever subsided (evanescent) since one year. On clinical examination patient had cervical and right inguinal lymphadenopathy with splenomegaly. Laboratory test showed anemia, neutrophilic leukocytosis, raised liver enzymes and hyperferritinemia. After excluding the all the potential causes for the patient's condition, she was diagnosed to have AOSD based on Yamaguchi criteria. On treating the patient with corticosteroids and NSAIDs patient improved symptomatically.

Keywords: AOSD: Adult - Onset Still's Disease

Introduction

Adult - onset still's disease (AOSD) first published by Bywater in 1971, is a rare multisystemic inflammatory disorder with an unknown etiology. Many theories have been put forward to understand the pathogenesis of the disease. The sign, symptoms and severity of the disease vary among different individuals. Spiking fever ≥39 °C, joint pain, skin rashes, and leukocytosis (≥10,000 cells/mm^3) have been described as the four main symptoms associated with it. AOSD has always been a diagnostic challenge, as many other clinical features other than the four common symptoms have been reported, with appearance of the disease in all age groups with potentially severe inflammatory onset [1].

Diagnosis is generally made after thorough clinical examination which requires exclusion of other conditions which mimics same presentation such as infection, malignancy and autoinflammatory conditions before labeling a patient to be affected with AOSD.

G. F. Still, described in children, 22 cases of chronic polyarthritis at Britain in 1897 which was known as Still's disease [2]. Children with systemic juvenile rheumatoid arthritis (JIA) who don't fulfill the criteria for classical rheumatoid arthritis have similar feature to AOSD.

The prevalence of AOSD is estimated to be one per 100,000 people. The age group of 15-25 and 36-46 years are generally affected with the disease [3].

Case History

A 42 years old female came to emergency department with complaints of low-grade intermittent fever for one year associated with non-pruritic rash predominately in lower limbs and trunk, arthralgia involving wrist, knee and ankle joints and with sore throat. On examination, she was pale, multiple cervical and inguinal lymphadenopathy with splenomegaly. Cardiovascular and respiratory system examination was normal.

Local causes, tuberculosis and lymphoma were ruled out following FNAC of lymph nodes which showed nonspecific reactive lymphadenitis. Further evaluation showed that she was anemic with Hemoglobin (Hb) of 8 gm/dl, total leucocyte counts (TLC) of 29000/mm^3 with neutrophil predominance and moderate elevation of aspartate aminotransferase (AST) of 157 IU/L and Alanine aminotransferase (ALT) of 98 IU/L. Acute phase reactants like c reactive protein was 41 mg/l. Serum ferritin was very high: 1053.8 ng/ml.

She had a history of loss of consciousness 6 months back for which she was diagnosed as aseptic meningitis and steroids was started following which patient improved symptomatically. But when she stopped steroid for past 2 months, relapse occurred. During that phase, liver biopsy was done suspecting autoimmune hepatitis but was negative.

ANA and autoimmune panel were negative too. Rheumatoid factor was also negative. Serum Immunoglobulin (IgG) level was very high : 1841 mg/dl. Computed tomography angiography ruled out Polyarteritis Nodosa.

After ruling out all possible causes, according to Yamaguchi criteria, patient was diagnosed with Adult Onset Still's Disease and started on Naproxen 200mg twice daily following which patient improved symptomatically within five days and was discharged from hospital with advice to follow up in Rheumatology OPD.

Discussion

Adult onset Still's disease (AOSD) is a rare multisystem inflammatory disease first described by Bywaters in 1971 [2,4]. Yamaguchi, established the diagnostic criteria which is commonly used for diagnosis of adult onset still's disease. The minimum of two major criteria and three minor criteria needs to be established to confirm the diagnosis against the Yamaguchi Criteria [5].

Yamaguchi criteria [5]		
Five or more criteria are required. Two or more criteria should be major.		
Major criteria	Minor criteria	Exclusion Criteria
Fever > 39°C lasting 7 days or longer	Sore throat	infections
Arthralgias or arthritis for 14 days or longer	Hepatomegaly or Splenomegaly	Malignancies
Typical Rash	Lymphadenopathy	Other Rheumatic disease
WBC count > 10,000/μL with >80% neutrophils	Abnormal amino-transferases	
	Negative rheumatoid factor and ANA	

Table 1

Theories have been postulated for the etiology of AOSD, one being Infective, though definitive agent hasn't been established. Patients with AOSD have been observed with elevation of Interleukin (IL)-1, IL-6, interferon (IFN)-γ, and Tumor necrotic factor (TNF-α) [6].

The early diagnosis can be very challenging for clinicians in many cases due to absence of typical rash which generally erupt during the febrile phase. C reactive protein and Sr ferritin are usually elevated. Almost three-quarters of AOSD patients have been seen with 5 times or above raised values of upper limit of serum ferritin and few have been recorded above 10 000 μg/L [7].

Conclusion

AOSD is diagnosis of exclusion. Characteristic clinical and laboratory findings are the basis of diagnosis. High index of suspicion is needed and patients improves with corticosteroids and non-steroidal anti- inflammatory drugs (NSAIDs).

Bibliography

1. Mitrovic S., *et al*. "Adult-Onset Still's Disease". *Periodic and Non-Periodic Fevers* 30 (2019): 93-132.

2. Bywaters EG. "Still's disease in the adult". *Annals of the Rheumatic Diseases* 30.2 (1971): 121-133.

3. Seung OP and Sulaiman W. "Adult-Onset Still's Disease: A Case Report". *Oman Medical Journal* 26.5 (2011): e022.

4. Magadur-Joly G., *et al*. "Epidemiology of adult Still's disease: estimate of the incidence by a retrospective study in west France". *Annals of the Rheumatic Diseases* 54.7 (1995): 587-590.

5. Yamaguchi M., *et al*. "Preliminary criteria for classification of adult Still's disease". *Journal of Rheumatology* 19.3 (1992): 424-430.

6. Efthimiou P and Georgy S. "Pathogenesis and management of adult-onset Still's disease". *Seminars in Arthritis and Rheumatism* 36.3 (2006): 144-152.

7. Kelly J., *et al*. "Sore throat and hyperferritinaemia". *Journal of the Royal Society of Medicine* 94.8 (2001): 400-401.