Case report

A CASE REPORT ON ADULT- ONSET STILL'S DISEASE

ABSTRACT

Background: Adult-onset Still's disease (AOSD) is an uncommon systemic inflammatory condition with an unknown cause, presenting with a set of three main symptoms: recurrent high fevers, joint pain (sometimes accompanied by arthritis), and evanescent skin rash.

Case report: We here describe a 38 years female patient presented to ER with complaints of fever on and off, body pain and swelling of multiple joints for 3 months and rash with itching all over the body for 1 month. Fever was associated with chills and rigors with negative rheumatological factor and infections. Investigations of bone marrow analysis revealed a normal cell count and a slight increase in eosinophils precursors, without any abnormal cells or parasites. Based on the Yamaguchi criteria, the patient was diagnosed with AOSD after ruling out cancer, infection, and rheumatic disease as potential causes. Future studies are required, more specifically to focus on the exact pathophysiology of AOSD.

INTRODUCTION

Adult-onset Still's disease (AOSD) is an uncommon systemic inflammatory condition with an unknown cause, presenting with a set of three main symptoms: recurrent high fevers, joint pain (sometimes accompanied by arthritis), and evanescent skin rash [1].

Systemic juvenile idiopathic arthritis (SJIA) and adult-onset still's disease (AOSD) are two distinct forms of the same illness, which vary in terms of age between 2-15 & 36-45 yrs.

In the 1970s, Eric Bywaters first used the term Adult-Onset Still's disease (AOSD) to describe an inflammatory condition that mostly affects young adults. The disease resembles childhood-onset Still's disease, now known as systemic-onset juvenile idiopathic arthritis (SOJIA), which was first reported by Sir John Still a century ago 2.

The incidence rate of this condition is approximately 0.16 per 1,00,000 cases. It tends to occur in two distinct age brackets, specifically between 15 to 25 years as well as 36 to 45 years 3.Both diseases are characterized by significant systemic inflammation and are associated with inappropriate activation of the innate immune system and overproduction of the proinflammatory proteins IL-1, IL-6 and IL-18 [4].

Since there is no specific test for the diagnosis of AOSD, YAMAGUCHI AND FUN classification criteria are used. According to Yamaguchi and Fautrel's diagnostic classifications, arthralgias are one of the key indicators [5].

Although regional guidelines have recently been developed in Italy and Japan, there is no internationally recognized standards for the management of AOSD.

We report a case of AOSD. In this case we discuss about diagnosis and management of adult onset still disease.

PRESENTATION OF CASE

A 38 years female patient presented to ER with complaints of fever on and off, body pain and swelling of multiple joints for 3 months and rash with itching all over the body Since 1 month. Fever was associated with chills and rigors. Patient was advised to get admitted for further evaluation and management.

On physical examination, the patient was pale, febrile, macular rash were present in upper half of body, left wrist joint was swollen and tender.

patient had visited multiple centers over period of 3 months. She also agrees that joint pain and fever had temporarily subsided when she was on NSAIDS. However due to lack of proper follow up in a particular centers and frequent use over the counter drugs, her symptoms failed to respond to that medication. Relevant investigations were initiated (TABLE 1).

TABLE 1- Initial blood investigations and Results

TESTS CBC	RESULTS
Hemoglobin	10.5 g/dl
Total Leucocyte count	14,500/mm3
Neutrophils	92%
Lymphocytes	7%
Eosinophils	1%
Platelets	3,22,000/mm3
ESR	80mm/hr
Peripheral blood smear	Normocytic normochromic anaemia
Total bilirubin	0.86 mg/dl
Direct bilirubin	0.47 mg/dl
ALT	33 U/L
ALP	495 U/L
Total protein	6.9 g/dl
Albumin	1.8 g/dl

A wrist joint x-ray was performed, revealing bone erosion and a reduced joint space. Her echocardiogram showed no abnormalities, but her serum ferritin level was significantly

elevated. Due to the suspicion of lymphoma based on radiological findings, the medical procedure of lymph node biopsy and bone marrow aspiration (BMA) was performed. The bone marrow analysis revealed a normal cell count and a slight increase in eosinophils precursors, without any abnormal cells or parasites.

Rightarmin biopsy showed mild leukocytoclastic vasculitis in the superficial dermal capillaries.

The investigations and clinical history that were obtained demonstrated that all the major and minor Yamaguchi criteria necessary for diagnosis AOSD were present. Further investigations were conducted in order to detect the existence of infections and rheumatic conditions (TABLE 2).

TABLE 2- Investigations for infections and rheumatic disorder

TESTS	RESULTS
Ferritin, Serum	3834 ng/ml
Iron, Serum	21 mcg/dl
Total Iron Binding Capacity, Serum	245 mcg/dl
Complement 3 (c3), Serum	217 mg/dl
Complement 4 (c4), Serum	36 mg/dl
Ebstein-Barr Virus	IgG : positive
	IgM : Equivocal
	Repeated IgM(after 2 weeks): Negative
Cytomegalovirus	IgG : Positive
	IgM : Negative
Rubella	IgG : Positive

	IgM : Negative

Based on the Yamaguchi criteria, the patient was diagnosed with AOSD after ruling out cancer, infection, and rheumatic disease as potential causes.

She was started on oral prednisolone 20 mg, blood transfusion was done and supportive care was given with analgesics and anti-pyretics. There was notable improvement of the rash and pain. Patient was discharged with T.Prednisolone 60 mg, T.Augmentin 625 mg, T.Ebast-DC, T.Acton-OR 1gm (SOS) and Mupirocin ointment.

DISCUSSION

Adult-onset still disease is a rare, distinct, and unrelated systemic autoinflammatory disorder that affects numerous genes throughout the body. The dysregulation of inflammation that results in a cytokine storm is the defining characteristic of AOSD [6].

Bella Y Mehta et al., (2019) states that clinical symptoms of AOSD are non-specific and can resemble hematological malignancies, rheumatological diseases, infectious diseases, and other rheumatological conditions. Spiking quotidian or double-quotidian fever, transient mild oligo-articular arthritis, and salmon colored macular or maculopapular eruptions are some of the disease's distinctive clinical manifestations.

Additionally, there have been reports of uncommon occurrences such as liver disease, pericarditis, pleural effusions, pulmonary infiltrates, and cardiac arrhythmias. Monophasic, intermittent, and chronic patterns are the main patterns in which the clinical course of AOSD can take place [7].

Although the exact cause of AOSD remains unknown, there are indications that multiple factors, such as genetic vulnerability, infectious triggers, inflammation activation, and inadequate resolution of inflammation, play a role in the development of AOSD was concluded by Meng-Yan Wang et al (2019)[8].

Stefan Vordenbaumen et al., (2022) determined that the Yamaguchi classification criteria can be used to determine the clinical diagnosis of AOSD because they have demonstrated good diagnostic accuracy when compared to other criteria. An AOSD activity score is being created by a working group of the European Alliance of Associations for Rheumatology

(EULAR). Alternative multidimensional scores, like the Pouchot "systemic score", omit significant complications like lung involvement and macrophage activation syndrome (MAS), and they haven't been sufficiently evaluated as a disease activity parameter on an individual basis in AOSD for clinical purposes[9].

The given treatment was according to the guidelines, "Evidence-based clinical practice guideline for adult Still's disease" by Toshihide Mimura (2018) et al., the patient seems to be improved without any further symptoms[10].

CONCLUSION

AOSD is an uncommon illness characterized by an unknown cause and process of developme nt. Patients who show symptoms of rash, arthritis, and fever should take into account the poss ibility of other diagnoses, such as malignancy, infection, and rheumatic disease, after excludi ng them as potential causes. Future studies are required, more specifically to focus on the exact pathophysiology of AOSD.

ABBREVIATIONS

AOSD	Adult Onset Still Disease
ALP	Alkaline Phosphatase
ALT	Alanine Transaminase
BMA	Bone Marrow Aspiration
CBC	Complete Blood Count
ER	Emergency Room
ESR	Erythrocyte Sedimentation Rate
EULAR	European Alliance of Association for Rheumatology
IL-1	Interleukin-1
IL-6	Interleukin-6
IL-8	Interleukin-18
MAS	Macrophage Activation Syndrome
NSAIDS	Non-steroidal Anti-inflammatory Drugs
SJIA	Systemic Juvenile Idiopathic Arthritis

SOS Sign of Symptoms

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