Research article

Clinical presentation of patients with adult onset still's disease in Nairobi: case series

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Abstract

Introduction: Adult Still's Disease (ASD) is a systemic inflammatory disorder of unknown etiology, typically characterized by a clinical triad (daily spiking high fevers, evanescent rash, arthritis), and a biological triad (hyperferritinemia, hyperleukocytosis with neutrophilia and abnormal liver function test).
Objective: This case series set out to describe the clinical characteristics of patients with ASD seen at a rheumatology clinic in Nairobi.
Results: After a record search, 8 patients were noted to have ASD. Fever and arthritis were noted to be most predominant presenting features with almost all the patients having hyperferritinemia.

Introduction

Adult Still's Disease (ASD) is a rare disorder, known to exist world-wide, with equal distribution between sexes, and with three quarters of the patients reporting disease onset between 16 and 35 years of age [1]. When starting before 16 years old, Still's disease is called Systemic-Onset Juvenile Idiopathic Arthritis (SOJIA), classified within the spectrum of juvenile idiopathic arthritis. No single cause for ASD has been identified, although a variety of infectious triggers and genetic factors have been suggested.

There are several epidemiological studies on ASD from around the world. A retrospective study of 62 patients from France estimated the incidence of ASD at 0.16 per 100,000 persons, with a bimodal peak at ages 15-25 and 36-46, and equal distribution between the sexes [2]. A retrospective study of 45 patients from the Netherlands reported a median age of onset of 25 years (range 16 to 65 years) with 27% presenting over the age of 35, and 60% women [3]. An epidemiological survey from Japan estimated the incidence among men at 0.22 per 100,000, and among women at 0.34 per 100.000, with a mean age of 38.1 years, 67% presenting over the age of 35, and 65-70% women [4]. However, several cases with onset of ASD after the age of 60 have been reported [5, 6].

The initial symptom of ASD is usually sudden onset of daily spiking high fever. Fevers typically peak once daily, in the late afternoon or early evening, generally exceeding 39°C and lasting under 4 hours, returning to normal in 80% of patients even without antipyretic treatment. Fever sometimes has a double quotidian pattern, with highest spikes occurring in the late afternoon or early evening. Overall incidence of fever in ASD across the largest retrospective studies is 96% [1, 7].

The classic rash in ASD is an evanescent salmon-pink, macular or maculopapular eruption, predominantly involving proximal limbs and trunk, which usually emerges with the fever, especially in the evenings .The rash can exhibit the Koebner phenomenon, and as a result may occur especially in areas subject to friction, i.e. tight clothing. The rash may be mildly pruritic, and is often confused with drug allergy. Histology shows mild perivascular inflammation of the superficial dermis, with primarily lymphocytes, histiocytes, and dermal edema. Immunohistochemistry may show C3 deposition in the blood vessel walls. Overall incidence of rash in ASD is 73%[1].

Musculoskeletal symptoms are found in the majority of patients with ASD. Arthritis may initially be mild, oligoarticular and transient, evolving over a period of several months into a more severe, destructive, symmetrical and polyarticular form. Most commonly affected joints are knees, wrists and ankles, although elbows, shoulders, hips, interphalangeal, metacarpophalangeal, metatarsophalangeal, and temporomandibular joints may also be involved [1,7]. Progressive changes in the wrist joint, with precipitate or carpometacarpal joint space narrowing, typically present 6 months after disease onset and may develop to ankylosis in 1.5 to 3 years. Incidence of arthritis in ASD ranges from 64% to 100% [1, 7].

Generalized myalgias, often coinciding with fever spikes are also found in the majority of patients. Myalgia may be severe and debilitating. Inflammatory myopathy is rarely found in ASD, but serum creatinine kinase and aldolase

concentrations can be slightly elevated [7]. Incidence of myalgias in ASD ranges from 56% to 84% [1, 7].

ASD has been associated with marked elevations in serum ferritin concentration in approximately 70% of patients [8]. Serum ferritin concentrations are usually higher in ASD than in other inflammatory conditions [8].

Case presentations

A total of 8 patients with ASD were identified after a record search of all patients at Nairobi rheumatology clinic. Their demographic and treatment variables were ascertained and are summarized in the Table 1.

Table 1: Demographic treatment and variables of the patients

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8
Age (years)	39	48	32	37	37	24	45	21
Gender M/F	M	M	F	M	F	M	M	F
Fever Y/N	Y	Y	Y	Y	Y	Y	Y	N
Arthritis Y/N	N	Y	Y	Y	Y	Y	Y	Y
Myalgia Y/N	Y	N	N	N	Y	N		N
Others (specify)						Spleno-megally		
Ferritin levels H/L	H	H	H	H	H	H	H	N
WBC H/N/L	N	N	N	N	H			N
Neutrophils H/L/N	N	N	N	N	H			N
LFTs N/AbN	N			N	N	ABN		N
ESR N/H		H	H	H	H	H		
CRP N/H		H		H			H	
RF P/N							N	N
ANA P/N							N	N
Other tests (specify)								

The age at presentation was between 21 to 48 years, with mean age of 35.4 years. There were 5 male patients. Fever and arthritis were the predominant clinical features with 7 out of 8 patients presenting with either features. One patient was noted to have splenomegally.

Laboratory parameters: Hyperferritinemia was seen in 7 patients. Only 1 patient had a high white blood cell and neutrophil count. Abnormality in liver function test was seen in 1 patient. A high acute phase response was seen in 6 patients, with 5 patients having elevated ESR, 3 with high CRP levels and 2 with elevations of both CRP and ESR. Where ANA and RF testing was available, the results were negative.

Discussion

This evaluation set out to review the clinical presentation of patients diagnosed with ASD based on the EULAR diagnostic criteria. Eight patients were noted to have ASD , after a records search of all patients attending rheumatology clinic in Nairobi. All the 8 patients had their clinical features reviewed in this evaluation. The mean age at presentation of the patients was 35.4 years, with an age range of 21 to 48 years. Five patients were male. These findings are consistent with several epidemiological studies on ASD from around the world. In the largest case series report from Africa, Cheikhrouhou Abdelmoula *et al*[9], studied 11 cases of patients with ASD with a similar mean age of 35.4 years. Data from 62 patients in France estimated the incidence of ASD at 0.16 per 100,000 persons, with a bimodal peak at ages 15-25 and 36-46, and equal distribution between the sexes [2]. A retrospective study of 45 patients from the Netherlands reported a median age of onset of 25 years (range 16 to 65 years) with 27% presenting over the age of 35, and 60% women [3]. An epidemiological survey from Japan estimated the incidence among men at 0.22 per 100,000, and among women at 0.34 per 100.000, with a mean age of 38.1 years, 67% presenting over the age of 35, and 65-70% women [4].

The commonest clinical feature was fever and arthritis (7 out of eight patients presenting with either presentation). Two patients (25%) had myalgia with one patient presenting with splenomegally. Several reports have noted fever to be the commonest clinical feature with some series reporting up to 96% occurrence of fever among patients with ASD [1,7]. Arthritis has also been noted in majority of patients, with incidence ranging from 64% to 100% [7]. Arthritis may initially be mild, progressing gradually to severe destructive forms in untreated patients. The evaluation was unable to determine the severity of the arthritis as some records were missing data on this.

Hyperferritinimea was noted in 88% of the patients. ESR was elevated in half the patients with CRP elevation reported in 38% of the patients. Abnormalities in liver function tests was only reported in one patient with only one patient having a high neutrophil count.

Conclusion

ASD is a rare disease in our local set up, but with more aggressive suspicion index and appropriate investigations more cases can be identified.

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