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Case Report Open Access

Adult Onset Still's Disease-Diagnosis on a Stand Still

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Abbreviations:

AOSD: Adult Onset Still's Disease; PUO: Pyrexia of Unknown Origin

Introduction

Adult-onset Still's disease is a systemic inflammatory disease, named after English physician Sir George Frederic Still (1861-1941) [1]. The classic presentation is the triad of persistent high spiking fever, joint pain and a distinctive salmon-colored rash (appearing typically during fever episodes). The symptoms are similar to other inflammatory diseases and to autoimmune diseases which do have characteristic antibodies, which must be ruled out with tests for those antibodies.

Prevalence is estimated at 1.5 cases per 100,000-1,000,000 population with a bimodal age distribution with one peak between 15-25 years and second between 25-36 years [2]. The etiology of adult-onset Still's disease is not known, but it presumably involves IL-1, IL-1β and IL-18 [3]. The diagnosis is clinical and not based upon serology. At least eight sets of diagnostic criteria have been devised [4-11], however the Yamaguchi criteria has the highest sensitivity [10].

Major criteria	Minor criteria
Fever of at least 39C for at least one week.	Sore throat
Arthralgia or arthritis for at least 2 weeks	lymphadenopathy
Non pruritic salmon coloured rash (usually over extremities while febrile).	Hepatomegaly or splenomegaly
Leucocytosis	Abnormal liver function tests
	Negative tests for ANA and RF

Table 1: Eight sets of diagnostic criteria for adult-onset Still's disease.

For diagnosis atleast 5 criteria are required with atleast 2 major. In our case all the major and 3 minor criteria were fulfilled. Though elevated serum ferritin levels is not yet considered as an criteria for diagnosis of AOSD in Yamaguchi criteria but there are several studies which has shown a strong association between extremely high serum ferritin (much higher than seen in any other infectious or autoimmune diseases) and AOSD [11-15] (Table 1).

Case Presentation

A 40 year female patient came with complaints of fever and joint pains since 1 month. The fever was high grade, intermittent with chills and rigors. It responded to antipyretic medication.

She had pain in multiple joint pains involving both the large and small joints. There was history of muscle aches as well. Patient also complained of rash appearing on the extremities during episodes of fever. There was no history of cough, breathlessness, loose motions, bleeding tendencies, abdominal pain, headache, and discharge from ears or eyes, burning micturition. There was no past history of similar episodes or any other major illness.

The patient consumed mixed diet and did not notice disturbances in sleep pattern. There was no significant medical, social and family history in the past.

Lab investigations showed leucocytosis with TLC of 16500/cu mm (predominant polymorphs) Serum ferritin was elevated at 3948 ng/ml (N R: 10-190). LFT, RFT, Serum Electrolytes, Serum Proteins were normal.

Patient was negative for Dengue IgM/IgG, Brucella IgM, Chikungunya IgM, Rapid Malaria test, Widal test, Weil Felix test, monospot and Paul Bunnell for infectious mononucleosis, VDRL, HIV and HbsAg. Chest X-ray and X-rays of involved joints and ultrasonography of abdomen were normal.

Thyroid function tests were normal. Repeated blood and urine cultures showed no growth.

Patient was RA/ASO negative. Patient was negative for rhematologic antibody panel namely anti-CCP, ANA, nRNP/Sm, SSa, SSb, Ro-52, Scl-70, PM-Scl, Jo-1, CENP-B, PCNA, dsDNA, Anti-histone, anti nucleosome, anti-ribosomal-P Protein and AMA-M2.

Skin biopsy was obtained from the lesion of left index finger which was suggestive of marked hyperkratosis, parakerotosis and acanthosis, the central portion showed necrosis of epidermis as well as fragmentation of collagen in superficial dermis and infilteration by neutrophils and eosinophils. The final impression was necrotizing inflammation due to small vessel vasculitis. Lymph node biopsy was done which showed reactive hyperplasia. In view of clinical presentation, lab investigation, negative antibody panel, skin biopsy

report and markedly elevated serum ferritin levels diagnosis of "Adult Onset Still's Disease" was made.

In view of the clinical presentation and examination findings, the infections and autoimmune disorders were considered as major differentials.

With help of serological tests the probable infections like enteric, brucella, infectious mononucleosis HIV disease and syphilis were ruled out, after which screening for autoimmune collagen vascular disorders was done which was negative.

Exclusion of two major (above mentioned) differentials directed us to proceed towards an alternate diagnosis.

Patient was treated with oral glucocorticoids and NSAIDS which were gradually tapered off and she responded well to the treatment with symptomatic improvement.

Conclusion

Adult onset still's disease is one of underreported and under diagnosed causes of PUO. Diagnosis is clinical and not based on serology. Extremely high serum ferritin levels is a peculiar characteristic of the disease (though the serum ferritin levels are elevated in many inflammatory, infections and malignant disorders but the levels in adult onset still's disease is much higher when compared to other above mentioned conditions). Hence we recommend serum ferritin levels to be included as diagnostic criteria for Adult onset still's disease.

In a case of PUO, consider adult onset still's disease as an important differential.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of written consent is available for review by Editor-in-Chief of this journal.

Authors Contributions

SB and AK analysed and interpreted the patient data and were the major contributors in writing the manuscript; VG and ZS contributed in analysing, writing and revising the manuscript.

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