

THE DIAGNOSTIC CHALLENGE OF ADULT-ONSET STILL'S DISEASEH. A. Rostamipour¹ and L. Rastegarian²¹Department of Internal Medicine, Jahrom University of Medical Sciences, Jahrom, Iran²Shiraz University of Medical Sciences, Shiraz, Iran

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ABSTRACT

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown etiology, characterized by symptoms including high fever, salmon-pink rash, and also inflammatory symptoms such as sore throat, arthritis or arthralgia and polyserositis [1,2]. The prevalence is around 1.5 cases per 100,000 - 1000,000 population. The age distribution of Still's disease has two peaks, the first one at the age of 15 - 25 years and the second one at 35 - 45 years [3]. The most sensitive diagnostic criteria is the Yamaguchi criteria [3]. At least 5 criteria are required for diagnosis, which should include at least 2 major ones. Elevated serum ferritin level is not yet considered as a criteria for diagnosis of AOSD in Yamaguchi criteria, however, there are several studies which have demonstrated a strong association between extremely high serum ferritin and AOSD [5]. This is a 44-year-old lady, who was admitted with the chief complaint of chills and fever since two weeks before admission. She complained of body pain, arthralgia, dry cough, sore throat and generalized maculopapular rash. She had a serum ferritin level of 9291 ng/ml. She was labeled AOSD.

Keywords: Adult Onset Still's Disease; Fever of Unknown Origin, serum ferritin**Abbreviations:** AOSD: Adult Onset Still's Disease; FUO: Fever of Unknown OriginAuthor Correspondence, e-mail: author@gmail.comdoi: <http://dx.doi.org/10.4314/jfas.v8i3.605>

1. INTRODUCTION

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown etiology, characterized by symptoms including high fever, salmon-pink rash, and also inflammatory symptoms such as sore throat, arthritis or arthralgia and polyserositis [1,2].

The prevalence is around 1.5 cases per 100,000 - 1000,000 population. The age distribution of Still's disease has two peaks, the first one at the age of 15 - 25 years and the second one at 35 - 45 years [3].

At least 7 sets of criteria have been suggested for the diagnosis of this disease. The most sensitive one is the Yamaguchi criteria [3]. Arthralgia is among the major criteria for diagnostic classifications (Yamaguchi and Fautrel).[4]

At least 5 criteria are required for diagnosis, which should include at least 2 major ones. In our case, all the major and two minor criteria were fulfilled [5].

Elevated serum ferritin level is not yet considered as a criteria for diagnosis of AOSD in Yamaguchi criteria, however, there are several studies which have demonstrated a strong association between extremely high serum ferritin (much higher than levels observed in any other infectious or autoimmune disease) and AOSD [5].

2. CASE PRESENTATION

This is a 44-year-old lady, who was admitted with the chief complaint of chills and fever since two weeks before admission. She had high grade fever associated with chills and sweating that responded to intravenous acetaminophen.

She complained of body pain, arthralgia, dry cough, sore throat and generalized maculopapular rash. The rash was richly colored during the episodes of chills and fever. She did not have dysuria, abdominal pain, diarrhea, vaginal discharge and sputum. She did not have any positive history of a significant illness.

The patient had a history of anxiety disorder 3 year ago. At the time of admission she complained from both knee and wrist pain and swelling and developed frank arthritis of both wrists in the course of hospital admission.

She had leukocytosis (22,000) with a shift to the left and bacteremia. Blood and urine cultures were negative. All serologic markers for rheumatologic diseases were negative. Procalcitonin was 1.0 ng/ml.

The patient work-up for infectious diseases was negative including malaria, Brucellosis, monospot test, HBS Ag, HCV Ab, HIV Ab and ASO titer. The titer of CMV IgG and IgM were 12.531 and 1.034, respectively. The skin test for mycobacterium tuberculosis was negative. She was admitted in the hospital for a work up of fever of unknown origin (FUO).

The patient underwent skin biopsy from the right leg, which showed nonspecific changes, but urticarial vasculitis could not be ruled out. Abdomino-pelvic and chest CT scan were normal.

Bone marrow aspiration and biopsy was done, which showed cellular marrow with moderate shift to the left in myeloid maturation accompanied with an increase in the number of promyelocytes.

Abdomino-pelvic ultrasound examination showed a small size liver hemangioma. Upper gastroduodenoscopy and colonoscopy were also done, which did not show any abnormal finding. The patient had a serum ferritin level of 9291 ng/ml.

The patient was labeled AOSD and received prednisolone 1 mg/kg and also methotrexate 15 mg weekly and was discharged from the hospital. Her fever and chills was stopped, and she did not develop any new skin lesions, but she suffered from arthralgia.

Leflunomide 20 mg daily was added to her medication. She improved gradually and did not have any complaint. She is under regular follow up.

3. DISCUSSION

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease of unknown etiology, characterized by symptoms including high fever, salmon-pink rash, and also inflammatory symptoms such as sore throat, arthritis or arthralgia and polyserositis [1,2].

The prevalence is around 1.5 cases per 100,000 - 1000,000 population. The age distribution of Still's disease has two peaks, the first one at the age of 15 - 25 years and the second one at 35 - 45 years [3]. The most sensitive diagnostic criteria is the Yamaguchi criteria [3].

At least 5 criteria are required for diagnosis, which should include at least 2 major ones. Elevated serum ferritin level is not yet considered as a criteria for diagnosis of AOSD in Yamaguchi criteria, however, there are several studies which have demonstrated a strong association between extremely high serum ferritin and AOSD [5]. Our patient had a serum ferritin level of 9291 ng/ml.

4. CONCLUSION

AOSD is a differential diagnosis for prolonged fever in adults. Our patient had Yamaguchi criteria (fever and chills, salmon colored rash, leukocytosis, sore throat and arthritis in the course of hospital admission) as well as high ferritin level.

It has been previously suggested that extremely high serum ferritin levels of more than 5,000 ng/L should be a main diagnosis criteria for AOSD [1]. We agree with this notion that high ferritin levels should be included among the diagnostic criteria of AOSD.

5. REFERENCES

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