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Case Report

Adult Onset Still's Disease in a Patient with Fever of Unknown Origin: A Rare Case

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Abstract

Adult onset Still's disease (AOSD) is a rare clinical entity with unknown etiology, characterized by arthritis, fever, evanescent rash, and other systemic presentations. AOSD generally does not overlap with other rheumatic diseases. AOSD is a diagnosis by exclusion, and it is one of the common causes of pyrexia of unknown origin (PUO) which can be life-threatening if mistreated. Although steroids are the first line of therapy, about 20%-30% of patients are refractory, intolerant, and/or relapse during tapering or upon discontinuation of steroids. There are no clinical guidelines in treating such patients, which is challenging.

Keywords: Still's disease, arthritis, fever, auto-inflammatory

INTRODUCTION

Adult onset Still's disease is an infrequent auto-inflammatory ailment with unclear etiology their impact is mostly seen in a young individual, hyperpyrexia arthritis and pink-colored rashes are the main manifestation of AOSD¹. The primary designs are monophasic, intermittent, and chronic distinguishing the analytical history of AOSD. The duration of monophasic ASOD normally ends from week to month but the majority of people experience full recovery in under a year². The initial line medication of AOSD is steroid³. This disease mostly impacts the age group of 15-25 and 36-46 years⁴. The 1st case of AOSD was declared in 1896⁵. In 1992 Yamaguchi criteria were used to identify AOSD that have a sensitivity of 96 and a specificity of 92. Four main standards of this are (i) pyrexia less than 39°C at least for a week (ii) arthritis that ends more than 2 weeks (iii) pink-colored rashes. Four minor Yamaguchi standards are (i) pharyngitis (ii) adenopathy and splenomegaly (iii) abnormal LFT finding (iv) RF test⁶. Anti-inflammatory treatment, immune suppressants, and rheumatologic medication (azathioprine, tacrolimus, or cyclosporine) are used in AOSD. Treatment for acute AOSD presentation may involve intravenous

immunoglobulins⁷. It has also been suggested that anti-TNF or interleukin treatments are adequate for treating AOSD⁸.

CASE REPORT

A 26 years old female patient was suffering from high grade fever which is above 102°F, body along with joint pain, cough, vomiting with general weakness since 1 month. The patient was admitted female medicine ward with these symptoms. No history of Diabetes mellitus type-2, or hypertension, but the patient was diagnosed with hypothyroidism where the TSH level is 9.56 uIU/mL, in the LFT test report alkaline phosphatase was very high 566 U/L (normal range 35-104 U/L). The patient was suffering from iron deficiency anemia, Hb was 7.2 along with a low iron level of 24.03 µg/dL along positive stool occult blood. TLC was 25 thousand. RF factor was 32 IU/ml (normal range <8 IU/ml). 24 hour urine protein 803.25 mg/24hrs (10-140) USG report showed mild hepatomegaly, HRCT showed bilateral pleural effusions, and moderate left pneumothorax was present along with increased LDH-lactate dehydrogenase 422.49 U/L 37°C (125-220). According to laboratory reports, the patient was diagnosed with adult onset still disease.

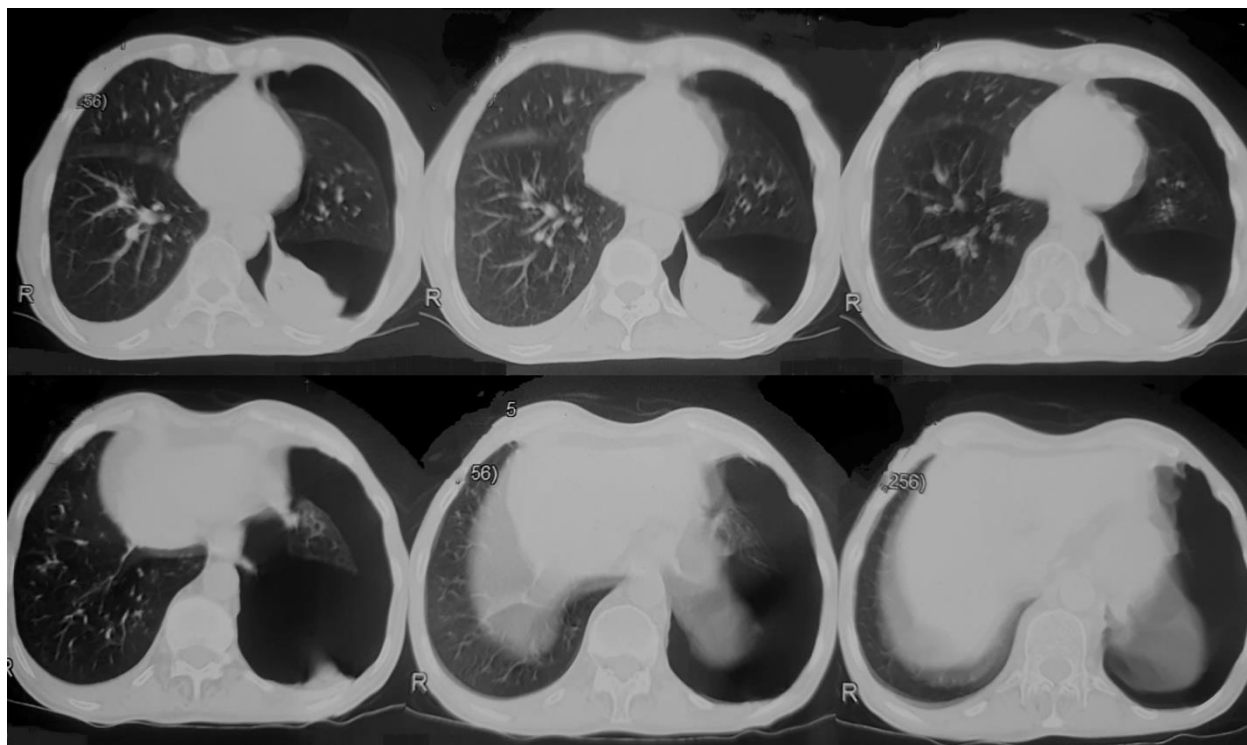


Figure 1: Chest HRCT report showed bilateral pleural effusions, and moderate left pneumothorax was present.

TREATMENT

The patient took medication according to the prescribed manner, tab. methotrexate 15mg once a week, tab. methylcobalamin 1500 mcg OD, tab. folic acid 5mg, tab. methylprednisolone 4 mg BD, tab. levothyroxine 75 mg OD, tab. hydroxychloroquine 200mg BD, tab calcium 500 mg vitamin D3 250 IU, patients take medicine continue 15 days. After 15 days the patient recovered slowly with the proper care of the healthcare team.

DISCUSSION

AOSD is a rare autoimmune disease that was first described in 1971. It is slightly more predominant in females with a ratio of 1:1.3 and the usual age of onset between 16 and 35 years; however, late onset of the disease is also possible and well known⁹. Common symptoms include high spiking fevers (typically 39 degrees Celsius or higher), evanescent rash, and polyarthritides. Multiorgan involvement with lymphadenopathy, liver dysfunction, elevated ferritin levels, sore throat, and hepatosplenomegaly can also be seen^{10,11}. Sore throat is one of the major signs of AOSD and may be associated withodynophagia¹². Arthralgia and arthritis mainly involving the knees, wrists, ankles and elbows have also been noted. The flare up of joint symptoms occurs during the febrile spikes^{13,14}. Carpal joints are the target of the most destructive arthritis in AOSD¹⁵.

The etiology of adult onset of Still's disease (AOSD) is unknown; both genetic factors and a variety of infectious triggers have been suggested as important, but there has been no proof of an infectious etiology, and the evidence supporting a role for genetic factors has been mixed. It is uncertain whether all patients with AOSD share the same etiopathogenic factors. Proposed pathogens have included numerous viruses; suspected bacterial pathogens include *Yersinia enterocolitica* and *Mycoplasma pneumoniae*¹⁶. The Yamaguchi criteria are widely used for the diagnosis of AOSD. When these criteria are used, it is necessary to exclude other rheumatic diseases; AOSD thus generally does not overlap with other rheumatic

diseases. Not much is known about AOSD as a co-morbidity with autoimmune diseases, including SLE. To the best of our knowledge, the present case is the first case of AOSD overlapping with SLE¹⁷.

CONCLUSION

Adult onset Still's disease is a rare condition with unknown etiology and pathogenesis, The diagnosis should be considered when patients show rash, arthritis, and fever after ruling out malignancies, infections, and rheumatoid conditions. As a result, not only will unnecessary tests be avoided, but also prompt treatment can be initiated in the form of steroids or biological inhibitors. It is an effective treatment option for patients with AOSD to use an anakinra injection of 2 mg/kg/day (maximum of 100 mg/day).

Declarations

Ethics approval and consent to participate

Informed consent was obtained from the patient for publication of this case report including the clinical information and accompanying images.

Consent for publication

The patient was assured that their name and initials will not be published.

Availability of data and material

This work has been carried out by us and we assure you that it can be provide to you whenever required.

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Conflict of interest

None declared.

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