

Adult Onset Still's Disease: A Case Report

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ABSTRACT

Adult onset Still's disease (AOSD) is a chronic multi-system inflammatory disorder characterized by high spiking fever, polyarthralgia and skin rash. Lymphadenopathy is another prominent feature of adult onset Still's disease. We describe a 22 years old lady presented with fever, skin rashes and polyarthritis for 3 months. Examination revealed fever, typical skin rash, generalized lymphadenopathy and polyarthritis. On investigation there were neutrophilic leukocytosis, high ESR, high ferritin level, but RA test and ANA test were negative. All of her history, clinical examinations and laboratory findings fulfill the diagnostic Yamaguchi criteria for AOSD. With proper treatment, now she is completely symptoms free and leaving a healthy life.

Key words: Fever, skin rash, polyarthritis, Adult onset Still's disease

Introduction

Adult onset Still's disease (AOSD) is a chronic systemic inflammatory disorder of unknown etiology, and its major clinical manifestations include high spiking fever, sore throat, muscle pain, polyarthralgia, salmon colored evanescent rash, hepatosplenomegaly, lymphadenopathy and neutrophilic leukocytosis^{1,2}. AOSD is rare and has a bimodal age distribution in all ethnic groups with peaks at 15-25 and 36-46 years of age in both sexes with an incidence of 0.16 cases/100000 persons/year^{1,2}. In fact there is no cure for adult Still's disease; however, treatment may offer symptom relief for adult Still's disease and help to prevent complication. As it progresses, adult Still's disease may lead to chronic arthritis and other complications. Recently we have diagnosed a case of AOSD in our hospital, who became complete symptoms free with proper treatment, though it is non curable.

Case Report

22 years of age normotensive, nondiabetic women admitted at department of Medicine, Dhaka Medical College Hospital with the complaints of fever, multiple joint pain and skin rashes over whole body for last 3 months. Fever was high grade continuous in nature. Generalized macular rashes especially in upper limbs, chest and back, with symmetrical polyarthritis involving wrist, proximal interphalangeal joints (PIP), metacarpophalangeal joints (MCP), knees and ankles. On quarry she also gave no history of sore throat for last 1 month. There were no early morning stiffness, ocular symptoms, orogenital ulcers, urinary symptoms, photophobia, and contact to infected person or major systemic symptoms.

Examination revealed well built, oriented young lady with macular rashes over upper limbs, chest and lower back and there was no rash over his finger knuckles. The patient was febrile 103°F with regular heart rate 90b/min, blood pressure of 110/70 mmHg and normal jugular venous pressure, but there were generalized lymphadenopathy involving bilateral anterior, posterior cervical groups and inguinal groups. She had acute synovitis of ankles, wrists and PIP joints, mainly of ring fingers with weak hand grip bilaterally, but full range of movements of all locomotor system and no proximal or distal muscular weakness. Deep tendon reflexes were normal. Examination of chest, abdomen, central and peripheral nervous systems were unremarkable.

Investigations revealed haemoglobin 7.5 gm/dl, erythrocyte sedimentation rate (ESR) 135 mm in 1st hour, total white blood cell count $12.17 \times 10^9/L$, neutrophil 67%, lymphocyte 14%, peripheral blood film suggestive of anaemia of chronic disorder with neutrophilic leukocytosis and marked rouleaux formation. Urine routine examination was normal, no growth in urine culture. Liver function test was normal, RA test - negative, ANA - negative, ultrasonography of the whole abdomen was normal, but there was very high ferritin - 4011 ng/ml (normal 15-120ng/ml).



Fig 1: Patient of AOSD with skin rashes

Finally AOSD diagnosis was made according to Yamaguachi criteria (she is having 4 major features and 3 minor features) and she was started on prednisolone 40mg daily and NSAID (Diclofenac sodium 100mg daily) after which she became afebrile for the first time in last 2 months after onset of illness. The patient showed considerable improvement and was discharged

home after one week on prednisolone 40mg daily with tapering dose of 5mg weekly, Diclofenac sodium and omeprazol 20mg twice daily. With proper treatment, now she is completely symptoms free and leaving a healthy life.

Discussion

Still's disease is named after an English doctor named George Still, who described the condition in children in 1897. George Still published his monograph, "On a Form of Chronic Joint Disease in Children."³ Still's disease is now known as systemic onset juvenile rheumatoid arthritis (JRA). Bywaters in 1971, the term "adult Still's disease" was used to describe adults who had a condition similar to systemic onset JRA². Although cause of AOSD is unknown, the condition may be triggered by a viral or bacterial infection.

High-sensitivity classification criteria have been proposed, since there is no single test to establish the diagnosis (Table 1)².

Table I: Classification criteria for adult-onset Still's disease proposed by Yamaguchi et al²

Major criteria	Minor criteria
• Temperature of $>39^{\circ}C$ for >1 wk.	• Sore throat
• Leukocytosis $>10\,000/cu\,mm$ and including 80% more of granulocytes.	• Lymph node enlargement
• Typical rash.	• Splenomegaly
• Arthralgias >2 wk.	• Liver dysfunction (high ALT)
	• Negative ANA, RF

After excluding infections, malignancies, and other rheumatic diseases, adult Still's should be considered if 5 criteria (2 of which being major ones) are present. ANA = antinuclear antibody; RF = rheumatoid factor. (Yamaguchi criteria 1992: specificity 92% and sensitivity 96%)

A salmon-pink bumpy or flat rash (87% cases) may come and go with the fever. The rash usually appears on trunk, arms or legs^{4,5}. Physical contact such as rubbing of skin may provoke the rash to appear. In 90% cases symmetrical or asymmetrical polyarthralgia or arthritis present, which usually involves knees, wrists, ankles, elbows, hands, shoulders joints and cervical spines^{4,5}. Usually, the joint discomfort

lasts at least two weeks. Lymphadenopathy is a prominent feature of AOSD seen in about 65% of patients and must do a biopsy to rule out lymphoma⁵. Life threatening conditions such as hepatic involvement, cardiac tamponade, disseminated intravascular coagulation (DIC), respiratory distress syndrome or pancytopenia were occasionally developed in the course of disease, and some cases were often associated with hemophagocytic syndrome (HS)⁶. Laboratory features of the disease are increased serum levels of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), leukocytosis, liver dysfunction, negative results for both rheumatoid factor and antinuclear antibodies, and an increased incidence of hyperferritinemia⁴. Increased serum ferritin level is a nonspecific finding and should not be regarded as a diagnostic test. That ferritin may be helpful for monitoring disease activity during treatment⁵.

Non-steroidal anti-inflammatory drugs (NSAIDs), such as aspirin, ibuprofen or naproxen, help to reduce inflammation⁵. People with high-fever spikes, severe joint symptoms or complications with their internal organs might require glucocorticoids, such as prednisone (0.5-1mg/kg/day)^{5,7}. Methotrexate has been used successfully in a small series of people to treat adult Still's disease. It may also be used as a "steroid-sparing agent," meaning that if one gives methotrexate, smaller doses of corticosteroids may be sufficient to control disease^{5,8}. Some patients are refractory to these conventional therapies. Tumor necrosis factor-alpha (TNF) blockers, include infliximab, adalimumab, etanercept, anti-interleukin-1, anti-interleukin-6 agents, and most recently anti-CD20-expressing B-cell antibodies are also effective in some cases, but no evidence of respond to other DMARD (gold, penicillamine). Other experimental drugs, including cyclosporine and anakinra, have also been successful in small groups of people^{2,5,7}. Even with treatment, it's difficult to predict the course of adult Still's disease. Some people might only experience a single episode, while for others adult Still's disease may develop occasional flair up or a chronic condition. About one-third of people with the disorder may fall into each of the above groups. Even if the patient is

symptoms free sometimes they need to continue medications to control inflammation and prevent complications. So, patient needs at least 6 months of treatment⁵.

Conclusion

AOSD is an important cause of pyrexia of unknown origin (PUO). It needs proper documentation of fever, exclusion of other diseases and observation for at least 6-8 weeks to diagnose the disease. But a definite diagnosis and proper managements can make the prognosis much better.

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