RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 12 | December 2015 | ISSN - 2249-555X

Adult Onset Stills Disease: Case Report

KEYWORDS

DR JAYESH DUTT	DR ASIF SHAIKH	DR MANASI SHAH
MD medicine professor at vs hospital, Ahmedabad	MD medicine third year resident in vs hospital; Ahmedabad	(MD medicine first year resident in vs hospital; Ahmedabad)

INTRODUCTION

Adult onset stills disease is a rare systemic inflammatory disorder of unknown etiology characterized by high spiking fever, evanescent salmon coloured rash, arthritis, and multiorgan involvement.

It is caused by aberrancies in the innate inflammatory pathways, and increased activation of IL-1beta is considered as a major event in their pathogenesis.

Diagnosis that is one of exclusion and differential diagnosis includes infection, neoplastic,and autoimmune disorders.

Although there are several set of classification but yamaguchi criteria present highest sensitivity (93%)

CASE REPORT

A 21 years old female patient, housewife , presented with the chief complains of high grade fever(>101F) with chills, intermittent in nature, associated with polyarthralgia for more than 15 to 20 days. Fever spike was accompanied with simultaneously appearing salmon pink rash over face and hands, associated with low backpain, bodyache, myalgias. associated with infertility. Medical and family history unremarkable. Not associated with history of oral ulcers, photosensitivity, sicca symptoms, redness of eyes or raynauds phenomenon. No history of palpable purpura or blurring of vision..no history of miscarriages.History of loss of weight (2kgs)in past 15 days

On examination, patient was febrile 103F(on admission),tachypneic with tachycardia, associated with moderate throat congestion, palpable cervical lymph nodes and pinkish rash over face maxillary area..per abdomen was soft and patient was conscious oriented with normal respiratory and cardiovascular examination..

Investigations:

Complete blood count revealed 22600 wbc with neutrophilia(85%) hb-9.13 and platelet count was 3.52

SGPT was 71 ALP was 276
PT INR APTT were normal

USG ABDO KUB was normal.
MP BY PARACHECK was negative
DENGUE IGM was negative
SERUM WIDAL was negative
2D ECHO, ASO CRP were normal, infective endocarditis or rheumatic heart pathology were ruled out.
RA factor was negative and ANA was negative- Rheumatoid arthritis and SLE were ruled out.

USG neck revealed enlarged lymph nodes; bilateral jugular chain, submandibular, cervical group of lymph nodes.

Blood c s had no growth.

ESR was 160 and CRP was 85.5

Serum ferritin was more than 2000, significantly high.

All the above investigations led to the most plausible diagnosis of adult onset still's disease; diagnosis of exclusion and all the Yamaguchi's criteria were satisfied and hence she was diagnosed with AOSD..

YAMAGUCHI'S CRITERIA FOR ADULT ONSET STILL'S DISEASE

Major criteria	Minor criteria	Exclusion criteria
Fever more than 39 degree Celsius intermittently for more than one week	Sore throat	(1)Infections (2)malignancies (3)inflammatory diseases
Arthralgia > 2 weeks	Lymphadenopathy and or splenomegaly	
Typical salmon pink rash	Abnormal liver function	
Wbc >10,000(>80% granulocytes)	RF,ANA negative	

Treatment:

Patient was treated with meropenem to treat neutrophilia, and later on when diagnosed with AOSD patient was treated with oral omnacortil(steroids) 1mg/kg for two weeks then taper up to 10 mg/day.. accompanied with sustained release indomthacin for symptomatic relief of pain and also as an anti-inflammatory. Patient showed marked improvement in a week..

DISCUSSION

AOSD is an autoimmune inflammatory multisystemic disorder of unknown etiology. It is associated with marked increase in production of interleukin 1 beta. .

A fever, equal or greater than 102 Fahrenheit, that comes on quickly once per day, usually in the afternoon or evening..mostly resolves without treatment.

Joint pain, warmth and swelling affecting a few joints at first –often knees and wrists, and then several joints. Morning joint stiffness often lasts for several hours..

A salmon pink colour skin rash that usually comes and

Volume : 5 | Issue : 12 | December 2015 | ISSN - 2249-555X

goes with the fever and usually doesn't itch. Flat spots or both flat spots and small raised bumps may appear on torso, upper arms or legs, or face.

Severe muscle aches may also ebb with fever,

A sore throat that may be severe constant and burning,

Other symptoms may include abdominal pain, pain while taking deep breath, swollen glands and unexplained weight loss.

AOSD develops most often in people before 45 yrs usually and is more common in females. Less than one in 1,00,00 people develop AOSD each year. It accounts for 10 to 20 % of all cases of JRA..

100% patients have high intermittent fever, 100% have joint inflammation and pain, 95% have faint salmon pink rash, 85 % have lymphadenopathy or splenomegaly, 85% have elevated WBC count, 60% have pleuritis or pericarditis, 40% have severe anemia and 20% have abdominal pain…

76% have onset before 35 years age and less than 5% have onset after age 40..

In this case… patient presents with rheumatological symptoms, like rashes, fever, arthralgia, infertility, yet all work up for autoimmune disorder ,infectios etiologies and neoplastic etiology were negative. With ferritin which was significantly high…. With yamaguchi criteria fulfilled… so diagnosis was adult onset stills disease…

Conclusion

AOSD is a rare autoimmune disorder. most characteristic presentation of condition is fever ,arthralgia or arthritis, sore throat, and skin rash. Fever of unknown origin is one of the most common presentation of AOSD. Diagnosis is based on exclusion of inflammatory ,autoimmune and neoplastic disease and no definite serologic marker is available at present.

REFERENCE 1. Bywaters EG. Still's disease in the adult. Ann Rheum Dis 1971;30:121–33. 2. Magadur-Joly G, Billaud E, Barrier JH, et al. Epidemiology of adult Still's disease: estimate of the incidence by a retrospective study in west France. Ann Rheum Dis 1995;54:587–90. 3. Crispin JC, Martinez-Banos D, Alcocer-Varela J. Adult-onset Still disease as the cause of fever of unknown origin. Medicine (Baltimore) 2005;84: 331–7. 4. Mert A, Ozaras R, Tabak F, et al. Fever of unknown origin: a review of 20 patients with adult-onset still's disease. Clin Rheumatol 2003;22:89 –93. 5. Pouchot J, Sampalis JS, Beaudet F, et al. Adult Still's disease: manifestations, disease course and outcome in 62 patients. Medicine (Baltimore) 1991;70:118–36. 6. Steffe LA, Cooke CL. Still's disease in a 70-year-old woman. JAMA 1983;249:2062–3. 7. Cagatay Y, Gul A, Cagatay A, et al. Adult-onset Still's disease. Int J Clin Pract 2009;63:1050 –5. 8. Petersdorf RG, Beeson PB. Fever of unexplained origin: report on 100 cases. Medicine (Baltimore) 1961;40:1-30. 9. Larson EB, Featherstone HJ, Petersdorf RG. Fever of undetermined origin: diagnosis and follow up of 105 cases 1970-1980. Medicine (Baltimore) 1982;61:269-92. 10. Bambery P, Kaur E, Bhusnurmath SR, Gupta A, Deodhar SD. Adult onset Still's disease in North India. A report on six patients. Rheumatol Int 1987;7:173-6. 11Singh S, Samant R, Joshi VR. Adult onset Still's disease: a study of 14 cases. Clin Rheumatol 2008;27:35-9. 12Uppal SS, Pande IR, Kumar A, Kailash S, Sekharan NG, Adya CM, et al. Adult onset Still's disease in northern India: comparison with juvenile onset Still's disease. Br J Rheumatol 1995;34:429- 34. 13.Efthimiou P, Paik PK, Bielory L. Diagnosis and management of adult onset Still's disease. Ann Rheum Dis 2006;65:564-72. 14.Mueller RB, Sheriff A. Scoring adult onset Still's disease. J Rheumatol 2010;37:2203-4.