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Case Report

Adult onset still's disease: a diagnostic dilemma

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ABSTRACT

Adult onset still's disease is a rare systemic inflammatory disorder of unknown aetiology, characterized by clinical triad (high spiking fever, evanescent rash and arthritis) and biological triad with lack of serological markers as a true gold standard makes diagnosis difficulty. Here is a case of 32year old male presented with high grade fever for 2 months, joint pains and swelling for 1month rash for 3 days. O/E: pallor and B/L tender, swollen ankle and knee joints, and P/A: splenomegaly. Investigations showed-Hb%. 8 gms, neutrophilic leucocytosis with thrombocytosis, ESR:72 mm/1h, CRP elevated. ASO-titre, RA factor and Anti CCP antibodies are negative. Adult onset still's disease is a heterogenous and rare systemic inflammatory disorder of unknown aetiology with lack of serological diagnostic modalities.

Keywords: Adult onset still's disease, Arthritis, Evanescent rash, High spiking fever, Pyrexia of unknown origin

INTRODUCTION

Adult-onset Still's disease is an inflammatory disorder that includes a triad of daily fever, arthritis, and rash. George Still first described the disease in 1896.[1] In other words, it can also be described as, systemic-onset juvenile idiopathic arthritis. This still's disease is most common in the pediatric population. This same condition is described as adult-onset still's disease when seen in adults, who are not fulfilling criteria for classic rheumatoid arthritis but having presentation similar to children with systemic-onset idiopathic juvenile arthritis.[2] This is a rare case report of a patient who presented with an unusual presentation and however we diagnosed the same condition

CASE REPORT

In present study 32 years old presented with high-grade fever for 8 weeks more of evening rise of temperature associated with chills, later he developed arthralgias mostly involving metatarsophalangeal joints (Figure 1) later involving the knee and shoulder joints for 1 month (Figure 2) partially subsiding with NSAIDs. He developed an evanescent rash over back that lasts for three days and disappears.

On examination at the time of admission patient had mild splenomegaly, cervical lymphadenopathy (<1 cm) were present. Investigations (Table 1) revealed normocytic normochromic anemia, neutrophilic leukocytosis with thrombocytosis, elevated ESR (72 mm/hr), normal renal function tests, liver function tests normal, normal uric acid levels, high CRP levels, CBNAAT negative, widal negative, QBC and smear for MP negative, dengue and chikungunya test negative, screening for HIV, Hep-b and C-negative, ANA levels normal, ANCA- negative, complete urine examination shows few pus cells, CXR, CT abdomen and chest negative for malignancy, X-ray of shoulder and foot shows arthritis features (Figure 1).

Table 1: Investigative profile of the patient.

Investigation	Lab value/ report
Haemoglobin	11 gm%
Total count	12700
Differential count	N-84, L-15, M-1
Platelets	2.5lakhs
ESR	72mm/hr
Blood urea	24mg/dl
creatinine	1mg/dl
Total bilirubin	1.6mg/dl
Direct	0.3mg/dl
SGOT	140IU/L
SGPT	72IU/L
Alkaline Po4	60IU/L
C- reactive protein	72mg/dl
Rheumatoid factor	Negative
Anti-CCP	Negative
ANA	Negative
ICTC	Negative
Hepatitis profile	Negative
Serum ferritin	13000ng/dl
USG abdomen	Mild splenomegaly
Dengue IgM ELISA	Negative
Smear for MP	Negative
Chikungunya antibodies	Negative
CBNAAT sputum	Negative
CT abdomen and chest	Negative for malignancy
Echocardiography	Normal

Table 2: Yamaguchi criteria.

Major Criteria
Fever >39 degrees lasting for more than 1 week Arthralgias or arthritis lasting for two or more weeks. Typical rash Leucocytosis >10000/mm with >80%bpolymorphonuclear cells Sore throat
Minor criteria
Recent development of lymphadenopathy Hepatomegaly or Splenomegaly Abnormal liver function tests Negative tests for antinuclear antibody and rheumatoid factor
Exclusion criteria
Infections Malignancies And other rheumatic diseases
Five or more are required with 2 or more being major criteria for diagnosis of AOSD

Fever does not subside even after the course of antibiotics. After consulting with rheumatologist, author go with serum ferritin levels, which seems to be very high (13000 levels), and the patient was diagnosed as AOSD after fulfilling "Yamaguchi criteria" (Table 2).[10]

Later he placed on aceclofenac 100 mg and prednisolone 1mg/kg and slowly tapered down over 4 months, however, the patient developed severe gastritis and reoccurrence of the symptoms he is shifted on to methotrexate 5 mg weekly, now patient is doing well on regular follow-ups.

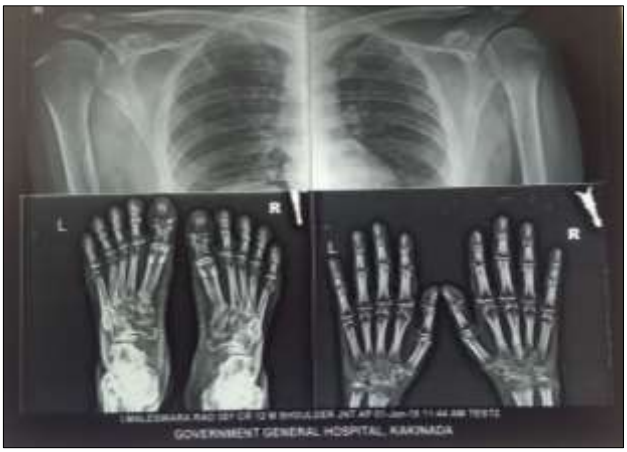

Figure 1: X-ray of shoulder, foot, hand showing osteopenia with no other changes.

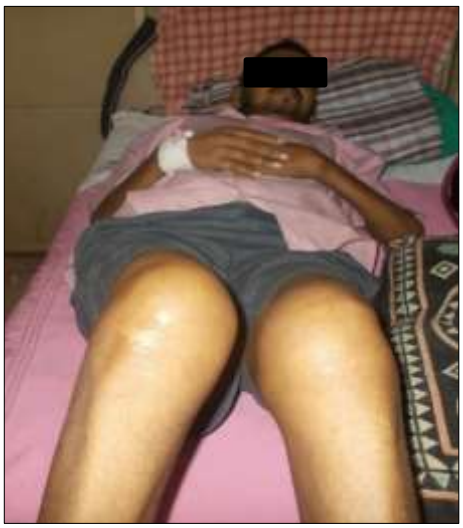

Figure 2: Bilateral swollen and tender knee joints at the time of admission.

DISCUSSION

AOSD is a rare systemic autoinflammatory disease with a prevalence ranging from 1 to 34 cases per million people.[2-4] Young people are preferentially affected with a mean age at diagnosis around 36 years old.[5,6] Clinical presentation is not specific, and a wide range of symptoms may be observed.[7] The actual etiology of still's disease is unknown. Though it is suggested infectious triggers can result in still's disease, there is no proven conclusive evidence. Some works of the literature suggested that autoimmunity mechanisms such as macrophage activation and inflammatory cytokines (e.g.,

IL-1, IL-6, IL-18, IFN-δ, and TNF-α), genetic factors, and infection may play a role of pathogenesis in AOSD.[8,9] Classification criteria used for adult onset still's disease known as Yamaguchi criteria (Table 2).[10] Fever, arthralgia, typical rash, and leukocytosis are major criteria for Yamaguchi whereas sore throat, lymphadenopathy, hepatosplenomegaly, abnormal liver function tests, negative antinuclear antibodies, and negative rheumatoid factor are considered minor ones.[8,9]

Other criteria used are Fautrel's criteria, spiking fever, arthralgia, transient erythema, pharyngitis, polymorphonuclear cells ≥80%, and glycosylated ferritin ≤20% are major criteria. Maculopapular rash and leukocytosis are minor criteria. Exclusion criteria was the presence of any infection, malignancy and rheumatic disorder are known to mimic should be excluded. Our patient in this case report fulfilled both Yamaguchi (Table 2) and Fautrel's criteria.[10]

The treatment generally relies on NSAIDs and steroids, requiring not furthermore potent immunosuppressors. In some refractory cases, hydroxychloroquine, gold salts, methotrexate and, cyclosporine have been used with limited data.

CONCLUSION

Thus to conclude, adult onset still's disease is a rare disease presented with nonspecific symptoms, so it should be kept in mind while investigating a case of PUO as there are no confirmatory test reports, but very high serum ferritin levels along with elevated ESR, high CRP show some specificity. This is more a kind of diagnostic exclusion, diagnosed with the help of Yamaguchi criteria.

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REFERENCES

1. Magadur-Joly G, Billaud E, Barrier JH, Pennec YL, Masson C, Renou P, et al. Epidemiology of adult Still's disease: estimate of the incidence by a retrospective study in west France. Annals Rheumatic Dis. 1995 Jul 1;54(7):587-90.
2. Masson C, Le Loët X, Lioté F, Renou P, Dubost JJ, Boissier MC, et al. Adult Still's disease: part I. Rev Rhum Engl Ed. 1995 Dec;62(11):748-57.
3. Wakai K, Ohta A, Tamakoshi A, Ohno Y, Kawamura T, Aoki R, et al. Estimated prevalence and incidence of adult Still's disease: findings by a nationwide epidemiological survey in Japan. J Epidemiol. 1997;7(4):221-5.
4. Evensen KJ, Nossent HC. Epidemiology and outcome of adult-onset Still's disease in Northern Norway. Scandinavian J Rheumatol. 2006 Jan 1;35(1):48-51.
5. Gerfaud-Valentin M, Maucort-Boulch D, Hot A, Iwaz J, Ninet J, Durieu I, et al. Adult-onset still disease: manifestations, treatment, outcome, and prognostic factors in 57 patients. Med. 2014 Mar;93(2):91-9.
6. Colina M, Zucchini W, Ciancio G, Orzincolo C, Trotta F, Govoni M. The evolution of adult-onset Still disease: an observational and comparative study in a cohort of 76 Italian patients. Semina Arthr Rheuma. 2011 Oct 1;41(2):279-85.
7. Ohta A, Yamaguchi M, Kaneoka H, Nagayoshi T, Hiida M. Adult Still's disease: review of 228 cases from the literature. J Rheumatol. 1987 Dec;14(6):1139-46.
8. Gerfaud-Valentin M, Jamilloux Y, Iwaz J, Seve P. Adult-onset Still's disease. Autoimmunity Reviews. 2014 Jul 1;13(7):708-22.
9. Yamamoto T. Cutaneous manifestations associated with adult-onset Still's disease: important diagnostic values. Rheumatol Inter. 2012 Aug 1;32(8):2233-7.
10. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, et al. Preliminary criteria for classification of adult Still's disease. J Rheumatol. 1992 Mar;19(3):424-30.

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