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Case Blog

Title: A Rare Presentation of Adult Onset Still's Disease

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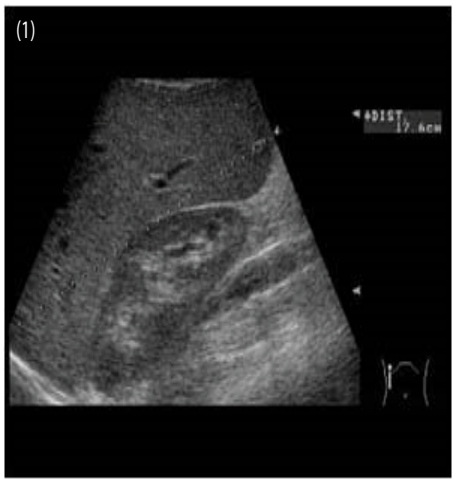

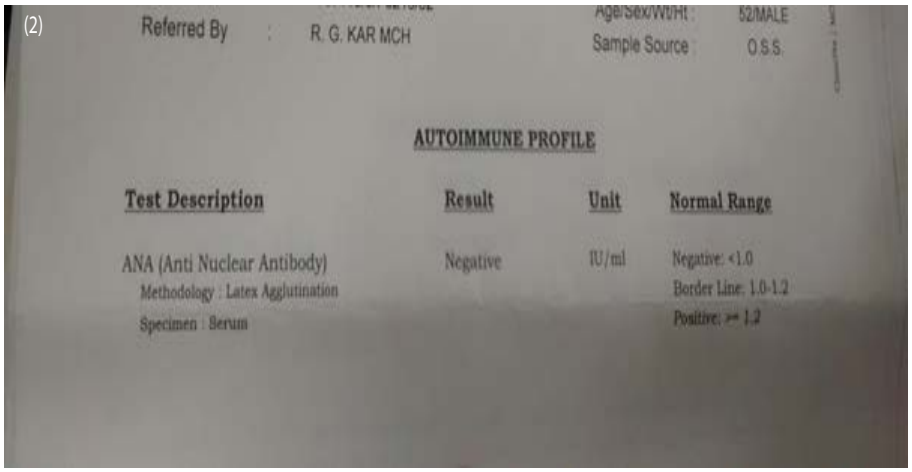

Figure 1: USG showing hepatomegaly.

Figure 2: Auto immune profiles showing negative ANA.

Introduction

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

Case Blog

A 52 year old male patient presented to ENT department with complaints of hyperplastic candidiasis of oral cavity. The patient was given management for candiasis, with oral and local antifungal. Simultaneously rapid ELISA was performed two at an interval of 6 weeks was done and results turned out to be negative. The candidiasis was not getting better even after a treatment of four weeks. After four weeks patient presented with history of arthritis of two weeks duration and fever for past seven days. Then patient was referred to medicine department for further workup. In medicine department routine blood investigations were done. Total count was twenty thousand nine hundred with eighty percent polymorphs. Erythrocyte sedimentation rate was one hundred and sixty seven mm/hour. Blood urea was ninety one, creatinine 3.5 mg/dl. Liver function test was showing normal bilirubin, SGOT 56 IU/L, SGPT 57 IU/L, alkaline phosphatase 149, total protein 5.3 gm/dl, albumin 2.9 gm/dl. Urine routine was found to be normal. Blood and urine culture and sensitivity revealed no growth. Ultrasound of the abdomen showed hepatospleenomegaly. Blood smear for malarial parasite came negative. HBsAg and anti HCV were non reactive. Chest X ray was within normal limits. to rule out associated macrophage activation syndrome bone marrow biopsy was done and the report showed reactive marrow with microcytic hypochromic RBC morphology.

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As patient had very high ESR with anaemia with azotaemia, to rule out multiple myeloma serum protein electrophoresis was done which elicited only low albumin with otherwise normal parameters and no M spike. To rule out the cause of anaemia iron profile was sent, which revealed serum iron of 98 microgram/dl. Serum ferritin was found to be 1563 ng/ml. transferrin saturation was 40%, reticulocyte count was 2.8%. Repeat ICTC for rapid ELISA came negative for HIV with normal CD4 counts. Blood for rheumatoid factor and anti nuclear antibody was found to be negative.

Thus according to the Yamaguchi criteria, factors for the diagnosis of Still's disease includes four major criteria and five minor criteria. To fulfil the diagnosis requires the presence of at least two major criteria. In our case five criteria was fulfilled including three being from the major criteria. Thus this case was concluded to be a case of adult onset Still's disease (Figure 1).

Discussion

The actual aetiology of still's disease is unknown. Though it is suggested that infectious triggers can result in still's disease, there is no proven conclusive evidence. Incidence of adult onset stills disease is very rare, in a study it is revealed that the incidence is 0.16 cases per one lakh population [2].

The classification criteria used for adult onset still's disease is known as yamguchi criteria [3]. The various sets of this criteria is as follows.

Major criteria

1. Fever >39 degree lasting for atleast one week.
2. Arthralgia lasting two weeks or longer
3. Maculo popular skin rash on trunk
4. Leucocytosis with atleast 80% granulocytes

Minor criteria

1. Sore throat
2. Lymphadenopathy
3. Hepatomegaly or splenomegaly
4. Abnormal liver function test
5. Negative test for antinuclear antibody and rheumatoid factor (Figure 2)

Exclusion criteria - the presence of any infection, malignancy or any other rheumatic disorder known to mimic Adult onset still disease, should be excluded in the diagnosis.

In our case, among the four major criteria three was positive and also among the minor criteria three were in favour of diagnosis.

The disease can follow three main patterns; they are as follows [4],

1. Monophasic pattern - disease course lasts weeks to months
2. Intermittent pattern - patient may have disease flares with or without articular symptoms
3. Chronic pattern - persistently active disease in which articular symptoms usually predominate.

The treatment of adult onset stills disease includes NSAID and steroid. Oral candidiasis, fever and arthritis subsided completely. After treatment total leukocyte count was 12,034, with 80% polymorphs. The patient was discharged after a period of six weeks. Till now there is no recurrence of the symptoms. The patient was followed up weekly for next 4 weeks, and once in 15 days for the next two months. Thus with the classification criteria this was a rare case report that presented with oral candidiasis, and diagnosed to be a case of adult onset still's disease.

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Conclusion

Thus to conclude, adult onset still's disease there is no confirmatory test reports. This is more a kind of diagnosis of exclusion, diagnosed with the guidance of Yamaguchi criteria. Thus it has to be kept in mind that sometimes rare presentation is also possible. This is one such presentation that presented with oral candidiasis and on further evaluation revealed Adult onset still's disease.

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