ADULT ONSET STILL’S DISEASE A CASE REPORT

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ABSTRACT

Adult Onset Still’s disease (AOSD) is a rare clinical entity characterised by fever, arthritis, and evanescent rash. This case report describes a 29 year old male who presented with classical features of AOSD diagnosed based on the Yamaguchi criteria.

KEYWORDS: Adult Onset, characterized, Yamaguchi criteria.

CASE REPORT

A 29 year old South Indian male, working for a Software firm presented with fever of 2 weeks’ duration. He also had evening rise of temperature. He did not have any other complaints. Examination revealed a well built male with temperature of 101°F with mild erythematous rash all over the trunk. Spleen was palpable. He did not have lymphadenopathy. There was no eschar. Hematological investigations revealed Total Count of 22,100 cells/mm³ with elevated ESR and CRP. All other reports including screening for Malaria, Dengue and other Viral haemorrhagic fevers were negative.

Chest X-ray was normal and Ultrasonographic examination of the Abdomen confirmed Splenomegaly. He was hence started on broad spectrum third generation Cephalosporins and empirical Doxycycline.

Even after 5 days of admission he continued to have fever and developed the rash during fever spikes. He also started developing polyarthritis involving both shoulders (maximum) both elbows and both knees causing painful restriction of joint mobility so much that he could hardly ambulate. Going by the protocols for workup of PUO, CT Chest and Abdomen were done which only revealed Splenomegaly. ANA, ds DNA, Anti CCP and RA factor were all negative. Retrovirus screening was also negative. Peripheral Smear Study done with great care revealed no abnormal cells.
His Serum Ferritin levels were markedly elevated at 16,500 µg/ml (Normal range 22 – 322 µg/ml) and ESR was 100 mm at 1 hour.

Based on his clinical features and laboratory evaluations, he was diagnosed to have AOSD using Yamaguchi Criteria. All intravenous antibiotics were stopped and he was started on Tab. Prednisolone 60mg (1mg / kg body weight) per day and NSAIDs for pain relief along with Calcium supplements and Proton Pump Inhibitors.

Patient showed dramatic improvement with the above management and was discharged after 1 week. Steroids were gradually tapered and stopped over the next 6-8 weeks. Repeat ferritin levels were 752 µg/ml and his glycemic status and bone density were normal at the end of treatment.

DISCUSSION
AOSD is a rare form of inflammatory arthritis first described by Eric Bywaters\(^1\) in 1971. Pathogenesis of this disease is still unclear. It has a bimodal age of distribution: 15 - 25 years and 36-46 years.

Patients with AOSD typically present with fever, arthralgia\(^2\) and sore throat.\(^5\) The rash is characterised by its Salmon pink colour. It is an evanescent non pruritic maculopapular rash usually present over trunk and extremities\(^3,4\) occurring during fever spikes\(^6,7\) Arthralgia and arthritis may involve major and minor joints as in our patient who had crippling arthritis.

Other features of AOSD not noted in this patient are: lymphadenopathy,\(^8\) pleuroperticarditis and CNS involvement.\(^9\)

Laboratory features include elevated ESR and marked elevation of Serum ferritin more than twice the upper limit of normal.\(^10\) Mild elevation of liver enzymes may be present.\(^11\) Rheumatoid factor and Antinuclear antibody are negative,\(^12\) as seen in our patient.

AOSD has close differential diagnosis especially in the early stages with other rheumatic diseases and malignancies also.

The Yamaguchi Criteria\(^13\) is the most widely used criteria for its diagnosis.

**Major Criteria**
1. Fever of at least 39°C for at least 1 week
2. Arthralgia or arthritis.
3. Evanescent non pruritic rash
4. Leukocytosis with granulocyte predominance.

**Minor Criteria**
1. Sore throat
2. Lymphadenopathy
3. Hepatomegaly / Splenomegaly
4. Abnormal LFT
5. Negative tests for ANA and RA.

Diagnosis requires at least five features with at least 2 major criteria. Our patient satisfied all the major criteria and had two minor criteria.

Non-steroidal anti-inflammatory agents can be used for pain relief.[14] Prednisolone[15] is given for patients not responding to NSAIDs at a dose of 1mg/kg body weight per day. Immunosuppressants may also be used in refractory cases.[16] Biologic agents are also used in rare cases.

The disease has a self limiting course of 6-8 weeks and Serum ferritin may be used to monitor response to treatment. Steroids are usually tapered and stopped after the acute episode.

Complications include joints destruction,[17] pericarditis, myocarditis etc. Patients may have recurrence of symptoms with chronic joint problems.

**CONCLUSION**
AOSD is a rare disease with unknown etiopathogenesis. It should be considered in adults presenting with rash, arthritis, fever after excluding other differential diagnosis discussed above.

**REFERENCES**