12. **MACROPHAGE ACTIVATION SYNDROME IN ADULT ONSET STILL'S DISEASE: A LIFE THREATENING COMPLICATION.**

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https://www.youtube.com/watch?v=0JIMP5Fyl7s&t=411s

INTRODUCTION: Adult onset Still's disease (AOSD), a multi-systemic inflammatory disorder, is a rare disorder but an important differential to be considered in patients of Pyrexia of Unknown Origin, especially if patient presents with polyarthritis. Macrophage activation syndrome (MAS), a subset of Secondary Hemophagocytic Lymphohistiocytosis (HLH), considered as the most life threatening complication of AOSD, mostly develops around the onset of disease. Hence, in a previously undiagnosed case of AOSD, recognizing MAS as a presenting feature complicating underlying AOSD is essential for increased patient survival, as in our case. **CASE:** A 39 year old diabetic female presented with high grade fever with chills and multiple joint pains symmetrically involving proximal shoulder, knees and distal joints (involving MCP, PIP and DIP joints of hand) associated with swelling and early morning stiffness, relieved on activity, since past 6 months. No history of oral ulcers, rash, jaundice, weight loss, loss of appetite, foreign travel or close animal contact. On examination, Pallor was present, Blood Pressure was 120/70 mm Hg, pulse rate was 102/min and temperature was 100 F. On per abdomen, mild hepatomegaly was present and other system examination was unremarkable. Routine laboratory findings on day of presentation have been summarised (Table 1.1). Ultrasound Abdomen revealed hepatomegaly and Chest X-ray showed signs of old infective foci. Peripheral smear for Malarial parasite, Ns1Ag and dengue serology, Widal test and Montoux test were negative. Blood cultures and urine cultures were sterile. X-Ray of joints were normal. USG of B/L Knee joints showed mild joint effusion bilaterally with no internal echoes (non tappable). Trans-thoracic echo and Trans-esophageal echo were normal. CT chest, neck and abdomen showed old tubercular changes in lung. Tests for atypical bacterial infections which was normal (RK 39, Chickungunya, Brucella, Leptospirosis and Scrub typhus). Fever did not respond to Broad Spectrum Antibiotics, Antimalarials or Antitubercular therapy and patient continued to have persistent fever spikes. Autoimmune profile showed ANA and RA Factor negative but markers of inflammation were raised: CRP- 521, ESR -148, IL-6 – 84.0, Procalcitonin-6.9 and S.Ferritin >2000. During the work up for Anemia, Microcytic hypochromic anemia was found with Elevated NAP SCORE-165. Incidentally, her triglycerides were found to be elevated (356) and a reduced fibrinogen. With these results, we planned for a bone marrow aspirate and biopsy which showed increased myeloid preponderance (32:1), increased histiocytes and evidence of hemophagocytosis. The 2004 diagnostic criteria of HLH was fulfilled. After ruling out almost all infectious and malignant causes of secondary HLH, we searched for a rheumatologic cause. Thus diagnosis of AOSD (after fulfilling Yamaguchi's criteria) with MAS (a subset of 2' HLH) was made. Patient had dramatic improvement after receiving steroids with her fever episodes and joint pains settling completely thereafter. **CONCLUSION**: Prompt recognition of life threatening complications like MAS which pose diagnostic difficulty due to overlapping features in a patient of AOSD, should be done at the earliest to improve patient prognosis and survival. Serum Ferritin levels can be considered a useful marker to assess the disease activity and to predict MAS occurrence in such patients.

Table. Lab Values on Day 1 of Presentation.

Lab Parameter	Value on presentation
Hb	**5.2**
TLC	**24260**
DLC	**92/7/1**
Platelet count	**5.9**
ESR	**141**
KFT	22/0.6
Na/K	138/4.1
Cal/Phosphate	7.9/3.4
Total Bil/D. Bil	0.76/0.16
ALT/AST	13/30
ALP	50
TPSA	6.5/3.0
CPK MB	27

Key words: Macrophage Activation syndrome; Adult Onset Still's disease; Hemophagocytic lymphohistyocytosis.