A Case of Macrophage Activating Syndrome Complicating New Onset Systemic Lupus Erythematosus

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INTRODUCTION

Macrophage activating syndrome (MAS) is a life-threatening disorder caused by inappropriate immune activation which may present as an acute febrile illness with lymphadenopathy, hepatosplenomegaly, and encephalopathy. Left untreated, MAS has a mortality of up to 50%[2] We present a case of MAS manifesting in the context of new-onset systemic lupus erythematosus (SLE).

CASE DESCRIPTION

A 26-year-old male with a family history of SLE presented with 3 months of generalized weakness, myalgias, swelling in his fingers and toes, and an unintentional 30-lb weight loss. Vitals on admission were remarkable for a temperature of 38.5°C. Exam was significant for soft, non-tender bilateral axillary and inguinal lymph nodes.

Initial Laboratory Evaluation:

WBC **2.66 k/uL** *(3.85 – 10.15)*
Hgb: **10 g/dL** *(13.3 – 16.2)*
Platelet: **103 k/uL** *(160 – 400)*

AST: **440 IU/L** *(7 – 40)*
ALT: **198 IU/L** *(10 – 60)*
ALP 153 IU/L *(60 – 350)*
Albumin 2.5 g/dL *(3.8 – 5.4)*

Ferritin: **>7500 ng/mL** *(24 – 336)*
Sedimentation Rate: **52 mm/hr** *(0 – 15)*
D-dimer: **3425 FEUng/mL** *(0 – 500)*
Triglycerides: **408 mg/dL** *(<175)*
hs Troponin I: **103.1 pg/mL** *(0 – 19.8)*

Subsequent right inguinal lymph node biopsy revealed reactive lymph node with follicular hyperplasia and broad areas of infarction without evidence of abnormal lymphoid cells on flow cytometry. A bone marrow biopsy showed normocellular marrow without evidence of hemophagocytosis or atypical lymphoid infiltrate. The diagnosis of MAS was made in the setting of fever, diffuse lymphadenopathy, splenomegaly, hypertriglyceridemia, pancytopenia, and elevated serum ferritin. He was started on intravenous methylprednisolone with marked clinical improvement and normalization of his inflammatory markers and was discharged with oral steroids and hydroxychloroquine.

INCLUSION CRITERIA – HLH 2004 Trial[1]

For diagnosis: Needing 5 out of 8 of the following:

Ferritin >500 ug/L	Fever >38.5 C
Splenomegaly	Bicytopenia (2 or 3 cell lineages)
Hypertriglyceridemia >2 mmol/L OR Hypofibrinogenemia <1.5 g/dL	Bone marrow biopsy demonstrating hemophagocytosis
Low or Absent NK Cell Activity	Soluble CD25 (IL-2) >2400 U/mL

Concurrent Rheumatologic Disorder
(ex. Systemic Juvenille Idiopathic Arthritis, Systemic Lupus Erythematosus)

MACROPHAGE ACTIVATING SYNDROME

IMAGES

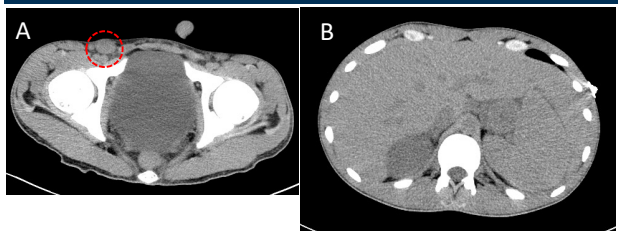

Fig 1 CT Abdomen / Pelvis Without Contrast. Images A and B. Image A demonstrating an enlarged right inguinal lymph node, with short axis measuring 15 mm. Image B demonstrating marked hepatosplenomegaly.

DISCUSSION

- This case demonstrates the importance of recognizing a life-threatening complication of new onset system lupus erythematosus. Macrophage activating syndrome is diagnosed when hemophagocytic lymphohistiocytosis presents in the context of a concurrent autoimmune disorder. This is most commonly systemic juvenile idiopathic arthritis.
- MAS is a potentially lethal complication in patients with autoimmune disorders with over 50% mortality in untreated cases.
- Early diagnosis is critical so that appropriate treatment with intravenous corticosteroids can be initiated to prevent tissue damage and death.

REFERENCES

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