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

Introduction

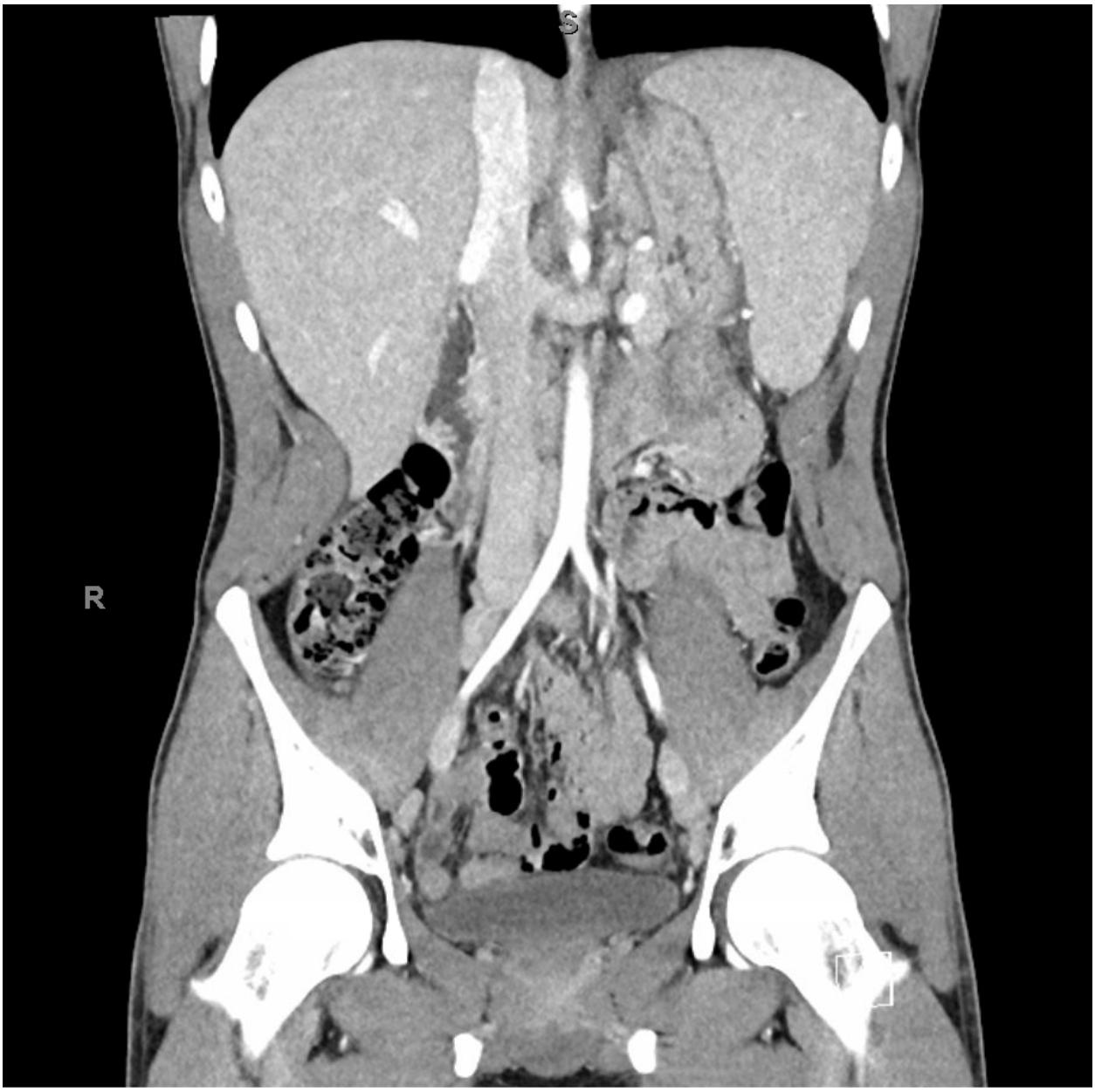
Macrophage activating syndrome (MAS) is a life-threatening disorder caused by inappropriate immune activation which may present as an acute febrile illness with lymphadenopathy, hepatosplenomegaly, and encephalopathy. We present an uncommon case of MAS manifesting in the context of new-onset systemic lupus erythematosus (SLE).

Case Presentation

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 were remarkable for a temperature of 38.5°C. His exam was significant for soft, non-tender bilateral axillary and inguinal lymph nodes. Blood count showed total white blood cells 2.66/mL, hemoglobin 10g/dL, and platelets 103/mL. Chemistry was significant for AST 440 IU/L, ALT 198 IU/L, ALP 153 IU/L, and albumin 2.8 mg/dL. His initial ferritin level was >7,500 ng/mL. Fibrinogen was elevated to 218/dL, and triglycerides 408 mg/dL. Erythrocyte sedimentation rate and C-reactive protein were negative. Rheumatologic evaluation established a diagnosis of SLE with positive ANA 1:640 with a homogenous pattern, anti-dsDNA 1:40, anti-Smith antibody >8.0, and low C3/C4 levels. Echocardiography obtained in the context of elevated troponin revealed an ejection fraction of 42.6% with a small pericardial effusion and moderate anterior wall hypokinesis consistent with lupus myopericarditis. CT of the abdomen and pelvis showed axillary and bilateral inguinal lymph nodes, and hepatosplenomegaly. 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.

Discussion

MAS is an uncommon but 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.



CT Scan of abdomen and pelvis showing enlarged liver, spleen and retroperoteneal lymph nodes.