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Lupus nephritis masquerading as hemophagocytic lymphohistiocytosis

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Hemophagocytic lymphohistiocytosis (HLH) is a rare, life threatening clinical syndrome characterized by hyperinflammatory cytokine storm due to exaggerated immune response. It may be triggered secondary to infections, malignancies, autoimmune diseases or medications. The following case report demonstrates acute lupus nephritis with initial presentation as HLH called acute lupus hemophagocytic syndrome. Estimated prevalence of HLH secondary to systemic lupus erythematosus (SLE) is rare between 0.9-4.6%. A 20-year-old African American female presented with progressive myalgia, malaise and recurrent fevers for 3 weeks. Vitals signs were normal except for temperature of 102°F. Her examination was unremarkable except for right posterior cervical lymphadenopathy. Laboratory data showed pancytopenia, hypertriglyceridemia, hyperferritenemia > 17,000 and hypoalbuminemia. Urine 24-hour protein was elevated > 5000 mg/24 hr. Bone marrow biopsy confirmed HLH and renal biopsy confirmed lupus nephritis. With the initiation of immunosuppressive regimen of dexamethasone and mycophenolate mofetil, she improved dramatically with resolution of fevers and normalization of HLH-specific disease markers. This case highlights the diagnostic challenge that may lead to delay in diagnosis of acute lupus hemophagocytic syndrome. Patients presenting with unexplained prolonged fever, cytopenias, abnormal liver function and elevated ferritin levels should prompt clinicians to perform immunologic testing for SLE in setting of HLH to avoid diagnostic and therapeutic delays.

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