Hemophagocytic Lymphohistiocytosis as Presenting Feature of Lupus

Hemophagocytic lymphohistiocytosis (HLH) is a multisystem disorder characterized by dysregulation of the immune system with hypercytokinemia and hyperinflammation. It can be primary or secondary [1]. Rheumatic diseases rarely cause secondary HLH. Systemic lupus erythromatosus (SLE) as a cause of HLH is rare [2-4].

A 7-year-old boy presented with fever of one month duration. On examination, he was febrile, liver and spleen were palpable 4 cms below costal margin. Initial investigations revealed a hemoglobin of 8.2 g/dL, TLC-1800/mm^3, Platelet-42,000/mm^3, MCV-94.6 fl, MCH-25.3 pg, MCHC-26.7g/dL. His peripheral smear revealed pancytopenia, macrocytes and microcytes. Serum widal titers were below 1:80. Blood culture and urine culture were sterile. Total protein was 4.8 g/dL, Albumin:globulin ratio was 0.5:1, Total bilirubin-2.7 mg/dL and were liver elevated enzymes (SGOT-180U/L, SGPT-513 U/L and ALP-570 U/L). Urea and creatinine were within normal range. Mantoux was non-reactive. He developed nasal bleeding and oral ulcers. Fever persisted inspite of treatment with ceftriaxone for seven days. Ultrasound of chest and abdomen revealed mild pericardial effusion, pleural effusion and mild ascites. Prothrombin time and activated prothrombin time were deranged, platelet counts were below 50,000/mm^3 and direct Coomb's test was negative. He received 4 unit of platelets and 2 unit of red cell concentrate transfusion within 10 days because of recurrent epistaxis, fall in hemoglobin and platelet count. Bone marrow aspiration revealed features suggestive of hemophagocytic syndrome (increase in histiocytes with hemophagocytosis). Triglyceride levels were 202 mg/dL. The diagnostic criteria of HLH syndrome were fulfilled [1]. In view of multisystem involvement with presence of pancytopenia, oral ulcers and serositis, diagnosis of SLE was considered. Antinuclear antibodies by ELISA method were positive. Anti-double stranded DNA levels were 169.9(>55 IU/mL significant). Child was diagnosed as having SLE as per the diagnostic criteria of SLE [5]. Renal biopsy did not reveal any abnormality. Child was given steroids (prednisolone 2 mg/kg/day). There was improvement in the clinical condition of the child, pleural effusion and ascites subsided. On follow up child did not have fever, bleeding manifestations, anemia or serositis. His hemoglobin was 11g/dL, TLC-7800/mm^3, and platelets-1.2 lakh/mm^3.

There are few case reports where SLE has manifested as HLH in a male child. Rajam, *et al*. [2] described two cases of HLH in rheumatic diseases in childhood. An adolescent girl with systemic onset juvenile idiopathic arthritis presented like severe sepsis and shock. The other patient presented with cardiac tamponade and was later diagnosed as SLE. Taki, *et al*. [3] reported a 21-year-old male diagnosed as SLE and HLH simultaneously. He was treated successfully with high-dose prednisolone. Yeap, *et al*. [4] described 14-year-old girl in whom HLH was the initial presentation of SLE. HLH as presenting feature of SLE in a male child is rare, as in our case.

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