Hematological abnormalities resistant to therapy in a child with systemic lupus erythematosus: Questions

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Case report

A 15-year-old girl diagnosed with systemic lupus erythematosus (SLE) 3 years ago with skin lesions, jaundice, fatigue, antinuclear antibody (ANA) and anti-double stranded DNA (anti-dsDNA), and complement consumption in other hospital. Elevated liver enzymes (AST 138 U/L -ALT 171 U/L) were observed. The serologies of the hepatotropic viruses (HAV, HBV, HCV) as well as the antibodies associated with autoimmune hepatitis (ASMA, anti-LKM1, anti-SLA, and anti-LP) were negative. Liver biopsy was performed and showed mild-to-moderate portal tract and limiting plate infiltrates consisting of lymphocytes and neutrophils with panlobular and porto-central necrosis. Acidophilic bodies were present. The findings were not compatible with autoimmune hepatitis or primary biliary cirrhosis. Considering the link between the liver enzyme abnormalities and biopsy results, and on exclusion of other causes, liver disease was considered lupus hepatitis. Her initial treatment included prednisone and hydroxychloroquine. During the following 2 years, the patient remained in stable clinical condition with elevated liver enzymes.

She was admitted to our clinic with oral aphthous lesions and bilateral knee pain and swelling. On physical examination, petechial lesions on the cheeks, dorsal side and upper extremities, and bilateral arthritis of the knees were observed. Her blood pressure was 110/75 mmHg. The remaining physical examination was unremarkable.

Laboratory analysis revealed a hemoglobin level of 9.1 g/dl, white blood cell count (WBC) 3400/mm³ with 89%

segmented neutrophils, 5% lymphocytes, and 6% monocytes. Platelets count was 35,000/mm³. Urinalysis and biochemical parameters except for hepatic enzymes (AST 49 IU/L, ALT: 97 IU/L) were in normal range. Her direct coombs' test was 1 (+). Erythrocyte sedimentation rate was 38 mm/h, C-reactive protein was 0.28 mg/dl (0–8 mg/dL). ANA was positive at a titer of 1/320, with C3 51.4 mg/dL, C4 10.4 mg/dL, and anti-dsDNA 19.4 IU/mL (normal < 10 IU/ml).

Blood cultures, serological tests, and molecular investigations for viruses (TORCH, HIV, HBV, and HCV serology) were taken. All microbiological tests were negative, except for CMV IgG titer that was positive at > 200 ng/mL (range 0–9). The urine and blood cultures showed no growth.

She was diagnosed as lupus flare because of hematological abnormalities, increased liver enzymes, positivity to autoantibody, and complement consumption. Pulse methylprednisolone (30 mg/kg/day, maximum 1 g/day) treatment was performed for 3 days and oral 2-mg/kg/day dose was continued. Mycophenolate mofetil was added to her treatment. Since hematological parameters did not improve, intravenous immunoglobulin therapy (1 g/kg/d for 2 days) and rituximab (375 mg/m²/dose) were given. However, hematological and biochemical parameters did not improve with the treatments.

Questions

1. What are the next steps that you would take to determine the cause of hematological abnormalities?
2. What is the diagnosis?
3. What should be the management in this patient?

Compliance with ethical standards

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