Hepatosplenic T-cell Lymphoma Presenting with Severe Hemolytic Anemia

Shahreen Billah, MD, Wei Feng, MD, Anthony Padula, MD, N. D. Nguyen, MD, Margaret Uthman, MD

a: Department of Pathology, University of Texas Medical School at Houston, Houston TX, 77030

Introduction:

Hepatosplenic T-cell lymphoma is a rare extra-nodal systemic neoplasm first described by Gaulard in 1986. It is derived from cytotoxic T-cell receptor gene rearrangement. It typically occurs in young adult men presenting with hepatosplenomegaly, marked thrombocytopenia and bone marrow involvement. We report a case with a review of literature.

Case Report:

This is a 30-year-old African American man who presented with severe hemolytic anemia. His laboratory findings included severe anemia with hemoglobin of 6.2 g/dL, 12% reticulocytes, platelet count of 193K/μL and microspherocytes identified on peripheral blood smear. Furthermore, the direct antiglobulin test (DAT) was negative and there was an elevated LDH (1609 U/L), and negative internal controls were performed with each antibody. Flow cytometry was performed on the liver biopsy and bone marrow aspirate. Cytogenetic studies were also performed on the bone marrow aspirate.

Material and Methods:

Fixed and paraffin embedded sections were stained with hematoxylin and eosin. Immunohistochemistry was performed on a representative section using the following antibodies: CD3, CD7, CD56, and MPO. Positive controls and negative internal controls were performed with each antibody. Flow cytometry was performed on the liver biopsy and bone marrow aspirate. Cytogenetic studies were also performed on the bone marrow aspirate.

Pathological Findings:

The liver biopsy showed a sinusoidal infiltration of medium-sized lymphocytes with scant to moderate cytoplasm and moderately condensed chromatin without prominent nucleoli (Fig 1-2). Immunohistochemistry and flow cytometry studies of the liver biopsy revealed the following immunophenotype: positive for CD3, CD7, and CD56; negative for CD20, TdT, MPO, CD4 and CD8.

Although the liver biopsy showed the classical findings for hepatosplenic T-cell lymphoma, the hypercellular bone marrow (Fig. 3-5) showed marked erythroid hyperplasia with a predominance of pronormoblasts (M:E ratio of 0.5) and the presence of a minor population (16%) of atypical lymphocytes which shared similar immunophenotype (CD3, CD7, and CD56 positive; CD5, CD4 and CD8 negative) as the liver infiltrate. No peripheral blood involvement was identified by flow cytometry studies. Cytogenetic studies of the bone marrow showed the following karyotype: 46, XY, inv(9)(p12q13).

References: