

Images in hematology-oncology

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A 38 year-old man with anemia and splenomegaly

CLINICAL HISTORY

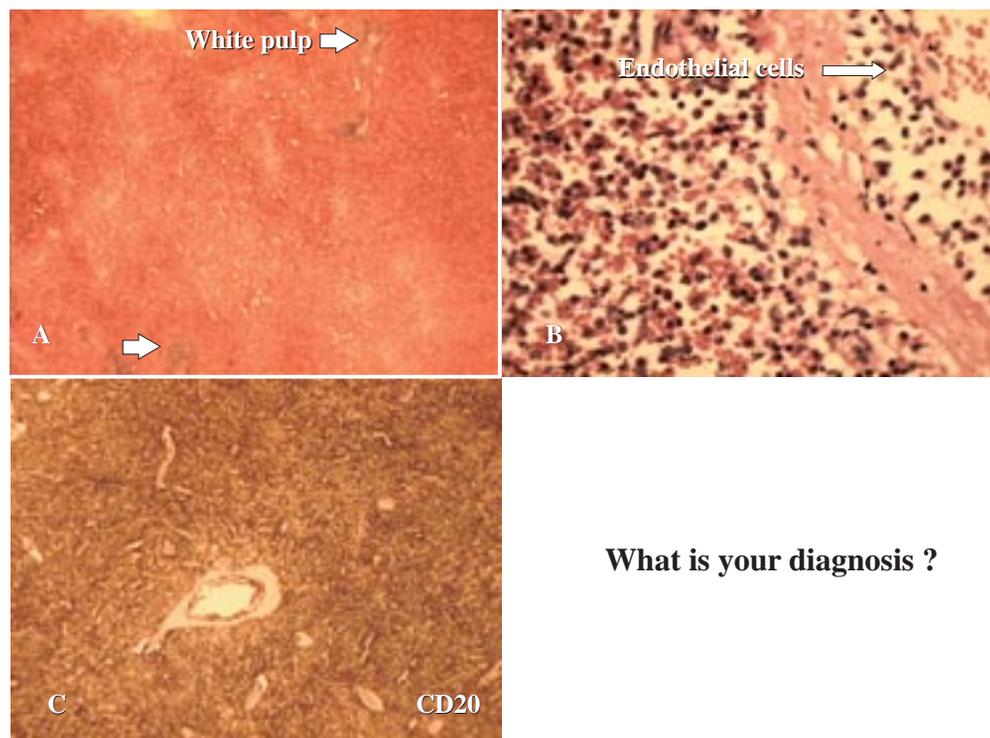
A 38 year-old man presented with the complaint of abdominal swelling and exhaustion. Splenomegaly was noted on physical examination and blood analysis revealed pancytopenia. Bone marrow biopsy was performed. On biopsy, the patient had a hypocellular bone-marrow with interstitial mononuclear cell infiltration which was CD20 positive. He underwent splenectomy.

On gross examination the splenectomy material weighed 570 grams and had a fairly smooth intact capsule. The cut surface was homogenous dark red with inconspicuous white pulp.

Microscopically there was diffuse infiltration of the red pulp with accompanying atrophy of the white pulp (Figure 1A). The diffuse infiltrate was composed of mononuclear cells with slightly pleomorphic round to oval nuclei displaying

homogenous chromatin pattern. Nucleoli were not present. The cells were widely spaced with barely discernable clear cytoplasm with prominent cytoplasmic borders. Focally there was subendothelial infiltration by these neoplastic cells (Figure 1B). Mitoses were absent. Focal areas of pooled erythrocytes could be found surrounded by the neoplastic cells (Figure 1B).

Histochemically the reticulin network was increased which could be seen surrounding individual neoplastic cells. Immunohistochemically these cells displayed CD20 (Figure 1C) and CD68 positivity. They were negative for cyclin D1 and bcl-2.



What is your diagnosis ?

PATHOLOGIC DIAGNOSIS

Hairy cell leukemia with splenic involvement

DISCUSSION

Hairy cell leukemia is a neoplasm of small B lymphocytes and derives its name from the irregular fine cytoplasmic projections of its individual cells, easily identifiable on smear preparations. It comprises 2% of lymphoid leukemias, typically presenting in middle aged males with massive splenomegaly and pancytopenia. The diagnosis is fairly straightforward when one encounters the typical mononuclear hairy cells characterized by an eccentric indented nucleus with spongy homogenous chromatin and pale slate-blue cytoplasm with long hairy projections on smear preparations (1,2).

Involvement of the spleen is fairly common in hairy cell leukemia and splenectomy is both a diagnostic and a therapeutic procedure. The diffuse red pulp infiltration is characteristic unlike the usual nodular infiltration of most lymphomas (1). The primary differential diagnoses for red pulp involvement in spleen is myeloid leukemias and large

granular lymphocytic leukemia. Mastocytosis and malignant histiocytosis are other neoplasms with similar diffuse red pulp involvement (3). The granular cytoplasm of mast cells can sometimes be appreciated in tissue sections and the pleomorphism of the cells of malignant histiocytosis is a helpful feature by itself. Red blood cell lakes surrounded by neoplastic cells and reticulin fibrosis is characteristic of HCL, aiding its differential diagnosis (1). Splenic lymphoma with villous lymphocytes enters the differential diagnosis especially on bone marrow aspiration and peripheral blood smears. They are, however, characterized by coarser chromatin pattern and shorter projections (3).

Immunohistochemistry greatly aids in confirming the diagnosis with hairy cells displaying CD20, TRAP and DBA-44 positivity. On immunohistochemical examination our case in addition to those mentioned also displayed DBA-44 reactivity which enabled us to confirm the already highly suspected diagnosis. TRAP by itself however is not diagnostic since it can also be present in SLVL. Cyclin D1 has also been shown to be present in HCL in addition to MCL (4,5).

References

- 1- Jaffe ES, Harris NL, Stein H. WHO Pathology and genetics: Tumours of hematopoietic and lymphoid tissues. IARC Press, Lyon 2001.
- 2- Allsup DJ, Cawley JC. The diagnosis and treatment of hairy-cell leukemia. *Blood Rev* 2002;16:255-62.
- 3- Wolf and Neiman. Disorders of the spleen. WB Saunders, 1989.
- 4- de Boer CJ, van Krieken JH, Schuurung E. Bcl-1/cyclin D1 in malignant lymphoma. *Ann Oncol* 1997;8:109-17.
- 5- Ishida F, Kitano K, Ichikawa N. Hairy cell leukemia with translocation (11;20)(q13;q11) and overexpression of cyclin D1. *Leuk Res* 1999;23:763-5.