Clinical and laboratory findings

A 65-year-old male patient was admitted who had been suffering for some months from weakness and sensory alterations of the legs, with difficulty in walking. At physical examination pallor, hepatomegaly and splenomegaly (6 cm below the costal margin) were found; there was no lymphadenopathy. Laboratory findings showed anemia (Hb 8.3 g/dL), leucocytosis (WBC 42.3 x 10^9/L), normal platelet count (234 x 10^9/L); serum paraprotein of the IgM λ type (60 g/L) and λ Bence Jones proteinuria.

Morphology

Eighty percent of peripheral blood cells had lymphoid features (Figure 1); they were larger than normal mature lymphocytes, had a round or oval nucleus, generally eccentric, sometimes apparently adhering to the cell membrane, condensed rather homogeneous chromatin, abundant blue cytoplasm with regular outlines and sometimes a paler-staining area adjacent to the nucleus (Figure 2). In a few cells more immature features were present such as open chromatin and nucleoli.

Bone marrow aspirate was easily obtained; lymphoid infiltration was small (10%) and many cells showed evidence of plasma cell differentiation. Histologic examination revealed that this infiltration had an interstitial pattern; there was no fibrosis.

Cytochemistry

Circulating cells were PAS negative and weakly positive to unspecific esterase and acid phosphatase; this last reaction was completely inhibited by tartaric acid.

Immunologic markers

From the immunologic point of view, most cells were positive to HLA-DR, CD5, CD19, CD20, CD22, CD38 and FMC7; they were negative to PCA-1, CD10 and T cell markers; SmIg (µλ) were expressed with medium-high density. In some cells light chain λ was also found in the cytoplasm.

Figure 1. Peripheral blood film showing many lymphoid cells and increased rouleaux formation. MGG x640.

Figure 2. Higher magnification shows cells with morphologic features intermediate between those of mature lymphocytes and those of plasma cells (lymphoplasmacytoid lymphocytes). MGG x1200.
Cytogenetics
Cytogenetic analysis evidenced the abnormal karyotype 48, XY, +3, +12 in a few mitoses.

Electron microscopy
Cytoplasmic features of plasma cells were found in elements with nuclear features of lymphocytes, i.e. many rough endoplasmic reticulum cisterns arranged in parallel (Figure 3). Cisterns were often disposed in concentric layers (Figure 4); the Golgi zone was well developed, with many electron-dense lysosomal granules nearby. Furthermore, mitochondria were morphologically atypical.

Conclusions
In conclusion, neoplastic cells had morphologic and immunophenotypic characteristics intermediate between those of mature B lymphocytes and those of plasma cells, and the patient had the clinical features of Waldenström's macroglobulinemia.

Electron microscopy confirmed the lymphoplasmacytic nature of the circulating cells.
Many chronic B cell lymphoproliferative disorders are characterized by a leukemic picture, as described in the review published by the FAB group.1-3 The leukemic phase of the splenomegalic variant of lymphoplasmacytic lymphoma has to be differentiated from other chronic splenomegalic leukemias; to accomplish this, besides the morphologic, immunophenotypic and possibly ultrastructural findings on peripheral blood cells, it is important to examine carefully the bone marrow, where morphologic features of plasma cell differentiation are often more evident than in peripheral blood.

References