

A VARIANT OF HAIRY CELL LEUKEMIA WITH UNUSUAL NUCLEAR MORPHOLOGY

Rosangela Invernizzi, Alessandro Castello*

Dipartimento di Medicina Interna, sezione di Clinica Medica II, and *Dipartimento di Patologia Umana ed Ereditaria, sezione di Anatomia ed Istologia Patologica, Università di Pavia, IRCCS Policlinico S. Matteo, Pavia, Italy

Clinical and laboratory findings

A 67-year-old man presented with progressive weakness, dyspnea and a three months history of daily intermittent fever. On physical examination the liver was slightly enlarged and the spleen was palpable 5 cms under the left costal margin. No lymphadenopathy was found. Complete blood count showed Hb 5.7 g/dL, platelet count $61 \times 10^9/L$ and WBC $2 \times 10^9/L$ with 35% neutrophils, 2% eosinophils, 60% lymphocytes, some atypical, and 3% monocytes.

Morphology

Atypical cells were larger than normal lymphocytes, had a moderate amount of agranular grey cytoplasm with irregular borders and fine hairlike processes; nuclei were irregular with prominent lobulation and convolution, finely reticular chromatin and sometimes small nucleoli (Figure 1).

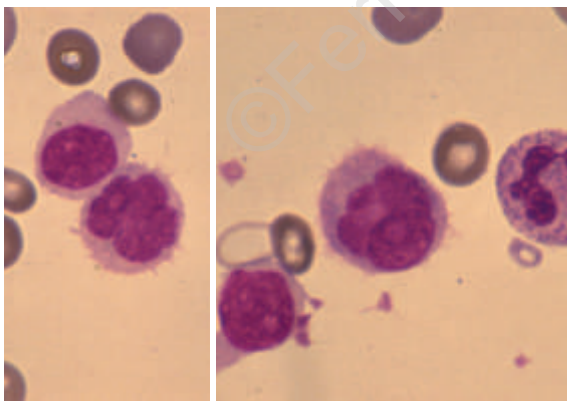


Figure 1. Peripheral blood leukemic cells show nuclear lobulation and convolution and fine hairlike cytoplasmic processes. MGG x1200.

Bone marrow aspiration gave *punctio sicca*; bone marrow trephine touch imprints showed infiltration by cells similar to circulating atypical cells with irregular nuclear outlines and round or ill-defined cytoplasmic borders (Figure 2).

Bone marrow trephine sections were hypercellular with almost complete replacement of normal cells by leukemic elements. The infiltration pattern was diffuse and at low magnification showed the aspect of hairy cell leukemia (HCL) (Figure 3a); it was loose and nuclei were widely separated by ample clear cytoplasm. The most striking feature of the cells was the very irregular nuclear shape (Figure 3b). There was a diffuse increase in reticulin fibrosis.

Cytochemistry and immunophenotyping

Leukemic cells were PAS and peroxidase negative, weakly positive to α -naphthyl-acetate esterase reaction and strongly positive with acid phosphatase after tartrate treatment with granules scattered throughout the cytoplasm.

Immunophenotyping of peripheral blood mononuclear cells showed that leukemic elements were of monoclonal B-cell lineage. They expressed monoclonal surface immunoglobulin of IgG λ type and were CD19, CD20 and CD22 positive; they expressed the activation antigens FMC7 and CD25, and also CD11c, but did not react with T-cell or myeloid monoclonal antibodies.

Conclusions

On the basis of these findings HCL with unusual nuclear morphologic features was

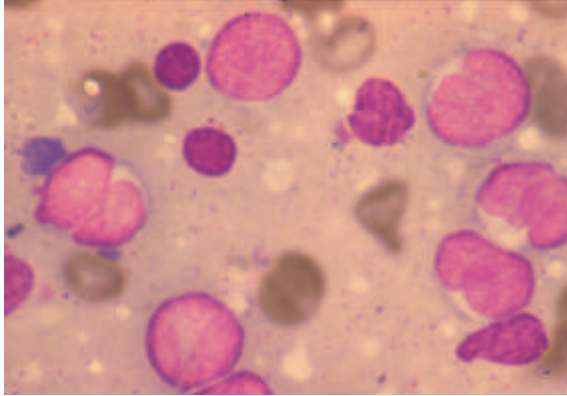


Figure 2. Leukemic cells from osteomyelobiopsy show irregular nuclear shape and round or ill-defined cytoplasmic borders. MGG x1200

diagnosed; therefore chemotherapy was begun with α -interferon.

Although HCL has generally been described as a homogeneous entity, rare morphologic and clinical variants have been recognized. Cawley et al. reported a variant showing features intermediate between HCL and prolymphocytic leukemia,^{1,2} the blastic form described by Martin et al. was characterized by leukemic elements similar to acute lymphoblastic leukemia blasts.³ More recently, a multilobular variant of HCL with morphologic similarities to T-cell lymphoma was observed.⁴ Many of the hairy cells of this form show marked nuclear lobulations and convolutions on both smears and tissue sections. Clinical, cytochemical and immunologic findings, identical to those of the classic HCL, are helpful in differentiating this

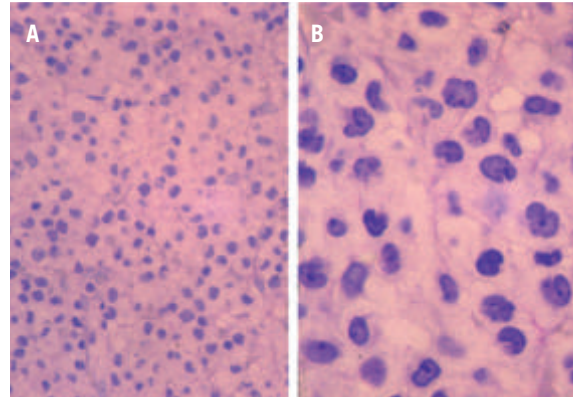


Figure 3. Bone marrow trephine section. Loose infiltration by cells with abundant cytoplasm and prominent nuclear irregularity. Plastic embedded, HE x120 (A), x500 (B).

variant from other chronic lymphoproliferative disorders with irregular or convoluted nuclei, such as peripheral T-cell lymphoma, Sezary syndrome, adult T-cell leukemia-lymphoma, follicular lymphoma and monocytoid B-cell lymphoma.

References

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