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Hairy cell leukemia with plasmacytoid morphology

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An 80-year-old man presented with persistent pancytopenia and splenomegaly, with a white blood cell count of 1.5× 10^9/L (normal range [NR] 4.0–10.0), absolute lymphocytes of 0.4 × 10^9 L (NR 1.2–3.7), hemoglobin and mean corpuscular volume of 10.8 g/dL (NR, 13.7–17.5) and 111.3 fL (NR, 79–92), respectively, and platelets of 71 × 10^9/L (NR, 150–330). Only exceedingly rare circulating lymphocytes

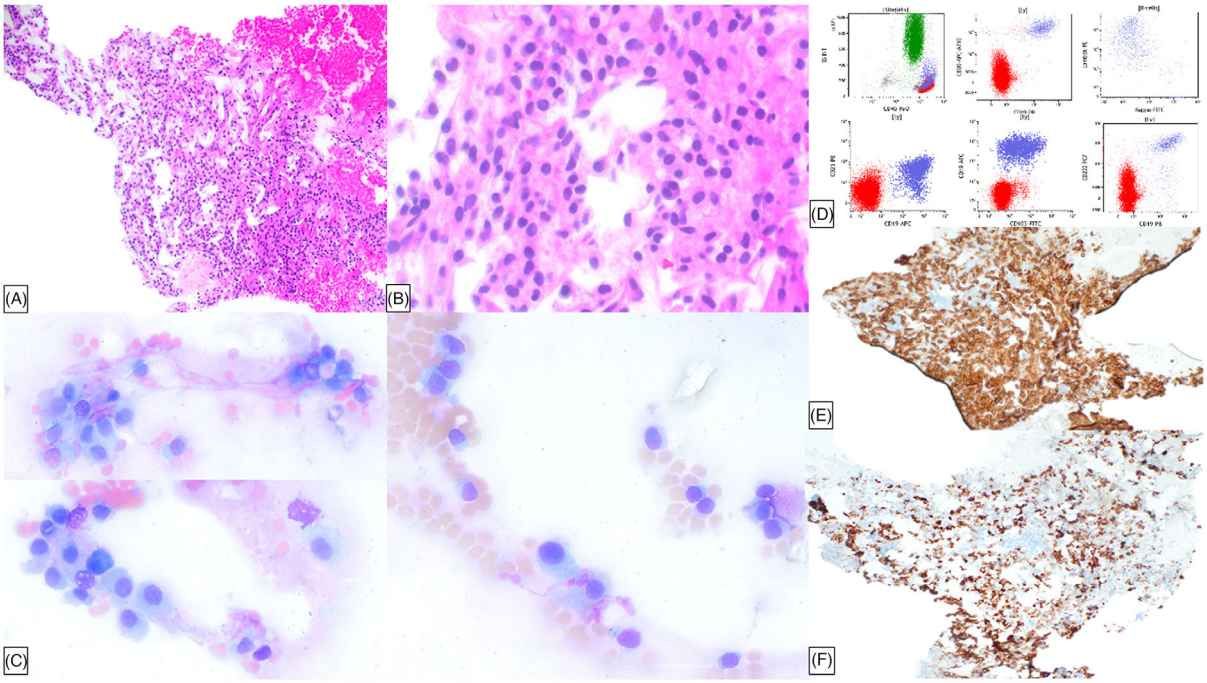

FIGURE 1 **Illustration of hairy cell leukemia with plasmacytoid morphology**: (A, B) Bone marrow core biopsy completely involved by a neoplastic infiltrate composed of plasmacytoid cells characterized by moderate to ample amount of pink cytoplasm & eccentric round nuclei; (C) Touch preparation of the core biopsy further highlights the plasmacytoid morphology of the neoplastic cells; (D) Flow cytometry analysis performed on marrow aspirate specimen demonstrates an aberrant B-cell population with an immunophenotype reminiscent of hairy cell leukemia: Bright CD20 expression, in addition to positivity for CD25, CD103, and CD200 expression; The neoplastic cells are positive for CD20 (E) and BRAF (F) stains by immunohistochemistry.

were detected on the peripheral blood smear, with moderate cytoplasm and eccentrically placed nuclei. A bone marrow biopsy was performed and revealed a hypercellular marrow for age (~40%) with decreased trilinear hematopoiesis and increased clusters of cells with plasmacytoid morphology **(A, B)** characterized by moderate to ample amount of cytoplasm & eccentric round nuclei on core biopsy **(B)** and touch preparation evaluation **(C)**, reminiscent of plasma cell neoplasm. Nevertheless, flow cytometry analysis performed on a hemodilute marrow aspirate demonstrated an aberrant B-cell population with an immunophenotype reminiscent of hairy cell leukemia (bright CD20 expression, in addition to positivity for CD25, CD103 and CD200 expression, with negativity of CD38 and CD138 expression) **(D)**. CD20 **(E)** and BRAF **(F)** stains highlighted the clusters of lymphocytes with plasmacytoid morphology on the core biopsy. No increased plasma cells were appreciated by stains or flow cytometry analysis (not shown). Next-generation sequencing performed on a hemodilute marrow aspirate detected a *BRAF* V600E mutation at a variant allele frequency of 2%.

This case illustrates a very rare and under-recognized morphologic presentation of hairy cell leukemia, which could be confused mainly with plasma cell neoplasm or lymphoma with plasma cell differentiation, such as lymphoplasmacytic lymphoma and marginal zone lymphoma. This morphologic variant of hairy cell leukemia expands the differential diagnosis of hematologic and non-hematologic neoplasms with plasmacytoid morphology that may involve the bone marrow. Adequate immunophenotypic evaluation is crucial in this setting to highlight the B-cell derivation of this neoplasm and to rule out other processes with plasmacytoid morphology (Figure 1).

AUTHOR CONTRIBUTIONS

Siba El Hussein conceptualized and wrote down the manuscript with valuable insights from Dennis P. O'Malley.

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The authors declare no conflict of interest.

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ETHICS STATEMENT

The information presented in this manuscript is deidentified, and there is no risk to the patient's privacy or confidentiality.

PATIENT CONSENT STATEMENT

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