

Exuberant nodal proliferation of mature plasmacytoid dendritic cells in a patient with chronic myelomonocytic leukemia



A 71-year-old man presented with an enlarged axillary lymph node 7 months after a diagnosis of chronic myelomonocytic leukemia 1 (CMML-1). Microscopically, the nodal architecture was nearly effaced by diffuse proliferation with scattered residual lymphoid aggregates (panel A; original magnification ×20, hematoxylin and eosin stain). The proliferation was composed of medium-sized cells with eccentrically located nuclei, irregular to indented nuclear contours, and relatively abundant eosinophilic cytoplasm without granules (panels B-C; original magnification ×400 [B] and ×1000 [C], hematoxylin and eosin stain). These cells exhibited the following characteristic immunophenotype: positive for CD123 (panel D; original magnification ×200), CD4 (panel E; original magnification ×200), CD68 (not shown), and TCL1 (panel F; original magnification ×200) with low Ki-67 (panel G; original magnification ×200), but negative for BCL2, CD2 (pink in panel H; CD123 in brown; original magnification ×200), CD3, CD5, CD19, CD34, CD56, CD68, lysozyme (pink in panel I; CD123 in brown; original magnification ×200), MPO, TdT, and TIA1 (data not shown). The overall features are consistent with a profound proliferation of mature plasmacytoid dendritic cells (PDCs) in association with CMML.

This case is consistent with the mature PDC proliferations associated with myeloid neoplasms, including, but not limited to, myelodysplastic and myeloproliferative disorders such as CMML. This mature PDC proliferation should be distinguished from the blastic PDC neoplasm recognized by the 2008 World Health Organization classification. The constellation of ample cytoplasm, low nuclear-to-cytoplasmic ratio, absence of CD56, and extremely low proliferation index distinguishes the mature PDC proliferation from the aggressive blastic PDC neoplasm.



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