

CD123⁺CD4⁺CD56⁺ neoplasm: blastic plasmacytoid dendritic cell neoplasm or acute myeloid leukemia?

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An 83-year-old-man with recent-onset multiple skin lesions had a bone marrow (BM) biopsy performed at an outside institution that showed increased CD123⁺CD4⁺CD56⁺ blasts suspicious for blastic plasmacytoid dendritic cell neoplasm (BPDCN). Complete blood count showed mild thrombocytopenia (platelets 130 \times 10⁹/L) and no circulating blasts. A repeat workup at our institution showed hypercellular BM with 35% blasts with oval nuclei, fine chromatin and moderate amphophilic cytoplasm (panels A-B; hematoxylin and eosin stain [A], Wright and Giemsa stain [B]; original magnification \times 500). Flow cytometry identified CD4⁺CD56⁺CD123⁺ blasts (panels C,F), which were CD13^{partial+}CD33⁺CD64⁺CD117^{partial+}HLA-DR⁺. Further workup showed blasts were butyrate esterase⁺lysozyme⁺CD11c⁺CD68⁺ and negative for CD14, TCL1, TCF4, CD303, MPO, E-cadherin, and B- and T-cell markers. Given the monoblastic morphology and

above immunophenotype, a diagnosis of acute myeloid leukemia (AML) was rendered. A skin biopsy showed extensive dermal infiltrate by immature mononuclear cells with an immunophenotype identical to blasts in BM consistent with leukemia cutis (panel D; hematoxylin and eosin stain, original magnification \times 200).

This case underscores the importance that clinical presentation of skin lesions and positivity for CD123, CD4, and CD56 on blasts can cause diagnostic challenge between BPDCN and AML. The finding that blasts were positive for monocytic markers (CD64, CD11c, lysozyme, and butyrate esterase [panel E; original magnification \times 500]) and negative for TCL1, TCF4, and CD303 was helpful in resolving this dilemma and correctly diagnosing AML. Differentiating BPDCN from AML is paramount, as therapy and prognosis differ significantly between them.



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