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Mature plasmacytoid dendritic cell proliferation in association with mixed-phenotype acute leukemia, T/myeloid

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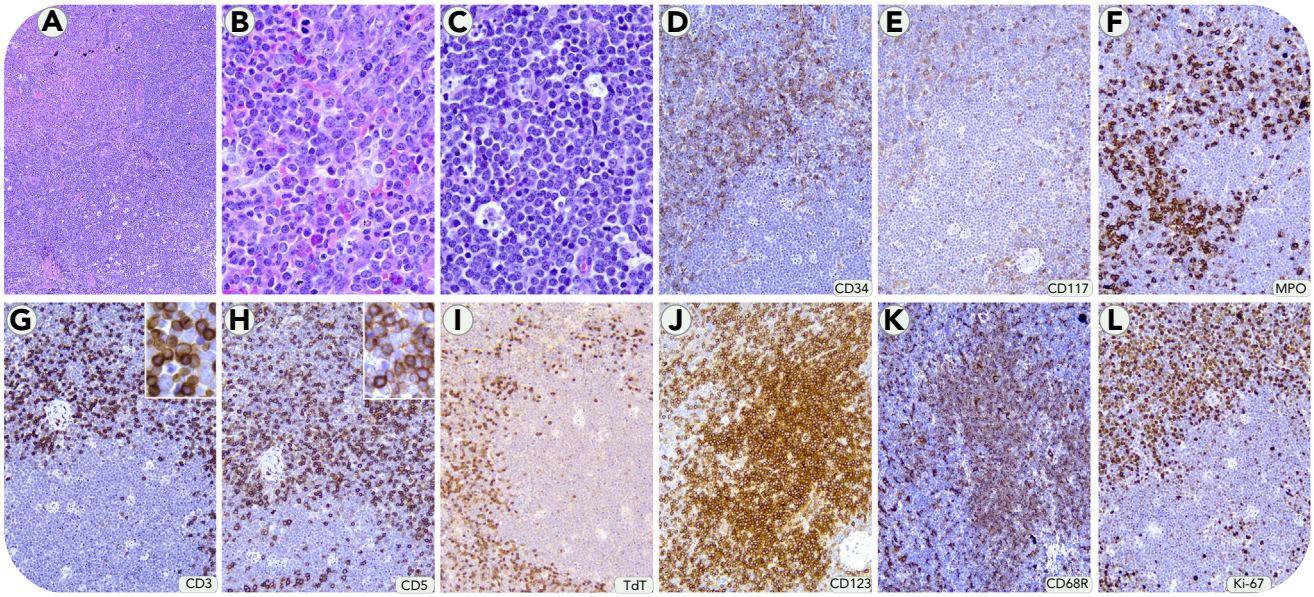

A 21-year-old woman presented with facial rash, leukocytosis (25.4 K/μL), anemia (7.8 g/dL), thrombocytopenia (40 K/μL), and circulating blasts. Bone marrow was hypercellular with ~20% myeloid blasts expressing CD34, CD117, CD33, lysozyme, and myeloperoxidase (MPO, subset) and lacking CD3, along with proliferation of plasmacytoid dendritic cells (pDCs) without CD56 and TCL1. Terminal deoxynucleotidyltransferase (TdT) was negative by flow cytometry. Cytogenetic and molecular studies revealed monosomy 7 without other common translocations and somatic variants. An excisional biopsy of the inguinal lymph node showed effacement of nodal architecture by blasts (panel A-B, hematoxylin and eosin stains, 20-40× objective) expressing myeloid and T-cell antigens at varying proportions, including CD3 (weaker than T cells), CD5 (weaker than T cells), CD34, CD117, MPO, and TdT (panel D-I, immunohistochemical stains, with hemotoxylin counterstains, 20× objective; panels G-H [insets], 60× objective). The pDC nodules with frequent apoptosis (panel A,C, 20-40× objective) had a similar phenotype to the marrow and showed expression of CD123 and CD68R (panel J-K, immunohistochemical stains, with hemotoxylin counterstains, 20× objective). Ki-67 proliferation index was 70% within the leukemic blasts and <10% within the pDC nodules (panel L, immunohistochemical stains, with hemotoxylin counterstains, 20× objective). The facial skin biopsy revealed infiltration by mature pDCs without leukemic blasts.

The clonal, nonmalignant proliferation of mature pDCs in association with immature neoplastic T cells in T/myeloid mixed phenotype acute leukemia is extremely rare. Leukemic cells demonstrate variable differentiation predilection at different anatomic sites, implying the importance of the microenvironment in leukemogenesis.

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