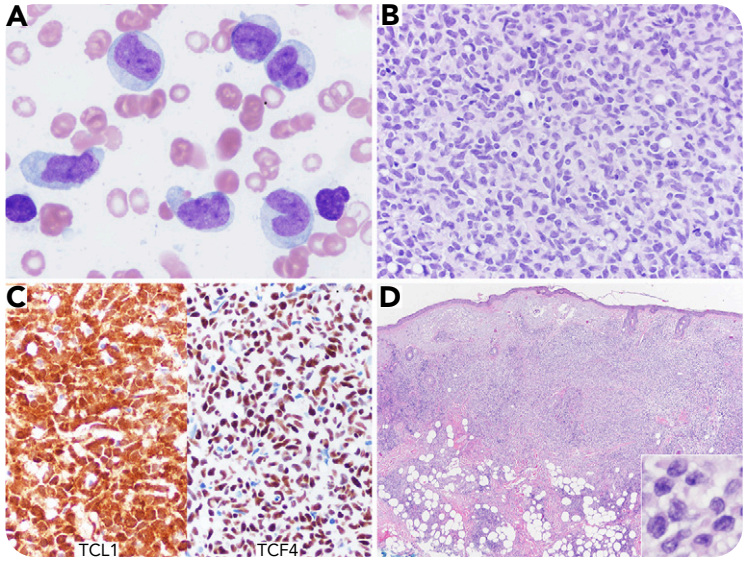
blood work
images in hematology

Acute leukemic variant of blastic plasmacytoid dendritic cell neoplasm at initial presentation

Hui Liu and Shimin Hu, The University of Texas MD Anderson Cancer Center



A 69-year-old woman presented with a 4-month history of low-grade fever, night sweats, 30-pound weight loss, and a skin lesion on the abdomen that was treated with antibiotics as a possible infection. At admission, her complete blood count was: white blood cells, 26.2 × 10^9/L (67% "blasts" with monocytic morphology); hemoglobin, 7.2 g/dL; and platelets, 11 × 10^9/L. A positron-emission tomography/computed tomography scan revealed extensive systemic hypermetabolic lymphadenopathies, as well as hepatosplenomegaly, pulmonary nodules, and diffuse bone marrow activities. A bone marrow specimen showed similar monocytic blasts (panel A; original magnification ×1000, Wright-Giemsa stain), replacing the entire medullary space (panel B; original magnification ×400, hematoxylin and eosin stain). The blasts in the peripheral blood and bone marrow were positive for CD2, CD4, CD7, CD33, CD38, CD56, CD123, CD303, HLA-DR, TCL1, and TCF4 (panel C; original magnification ×400) and negative for myeloid (CD13, CD15, CD117, and MPO) and monocytic markers (CD11c, CD14, CD64, and lysozyme). A left cheek skin biopsy revealed a tumor with a similar morphology and immunophenotype (panel D; original magnification ×40, insert, original magnification ×1000, hematoxylin and eosin stain). A diagnosis of blastic plasmacytoid dendritic cell neoplasm was established. She was started on hyper–cyclophosphamide, vincristine, doxorubicin, and dexamethasone plus venetoclax.

The uncommon features of this case include that the tumor presents as acute leukemia that occurs rarely at initial presentation, morphologically mimics acute monocytic leukemia, and demonstrates extensive extramedullary and medullary involvement without a concurrent myeloid neoplasm.

DOI 10.1182/blood.2020005039