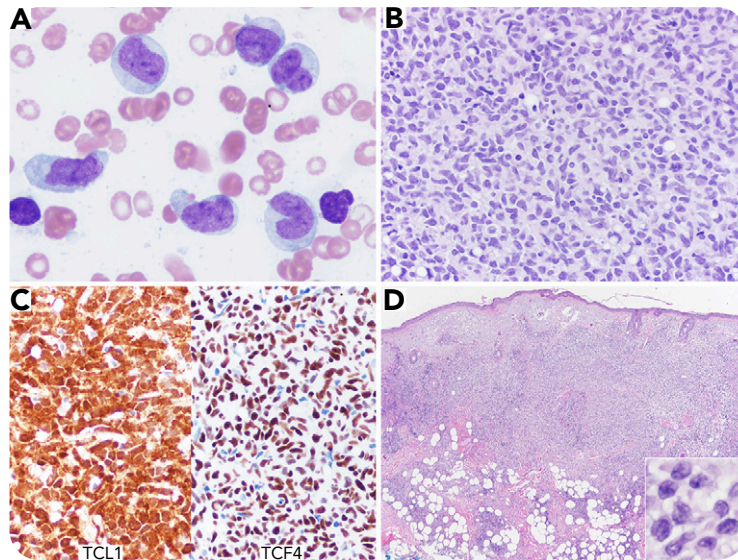


Acute leukemic variant of blastic plasmacytoid dendritic cell neoplasm at initial presentation

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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, $26.2 \times 10^9/L$ (67% "blasts" with monocytic morphology); hemoglobin, 7.2 g/dL; and platelets, $11 \times 10^9/L$. 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 $\times 1000$, Wright-Giemsa stain), replacing the entire medullary space (panel B; original magnification $\times 400$, 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 $\times 400$) 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 $\times 40$, insert, original magnification $\times 1000$, 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.