A 64-year-old man presented with progressive skin lesions, which were diagnosed as blastic plasmacytoid dendritic cell neoplasm (BPDCN) and were confirmed with marrow involvement. He received SL-401 chemotherapy but developed persistent monocytosis. A 3-month follow-up bone marrow biopsy showed characteristic “tadpole” cells on smears (panels A-B; original magnifications ×500, hematoxylin and eosin stain [A] and ×1000, Wright/Giemsa stain [B]) and patchy neoplastic cells positive for TCL-1 (panel C; original magnification ×500, TCL-1 immunostain), CD123, and CD4 (panel D; original magnification ×500, CD4 immunostain) by immunohistochemistry. Flow cytometry analysis revealed 2 aberrant populations (panel E; B, BPDCN; M, chronic myelomonocytic leukemia [CMML]), with dimmer CD56 in BPDCN population after treatment. Next-generation sequencing of the marrow aspirate demonstrated TET2 mutations. A diagnosis of BPDCN with CMML was made. Despite chemotherapy, his CMML progressed to acute myelomonocytic leukemia, and magnetic resonance imaging indicated leptomeningeal disease with spinal fluid showing numerous malignant hematopoietic cells. He died 10 months after BPDCN diagnosis.

BPDCN is an aggressive neoplasm that can be associated with other myeloid malignancies. Its association with CMML is of particular interest because CMML cases may also contain plasmacytoid dendritic cell nodules. Our case demonstrated immunophenotypic correlation between these 2 entities: both share expressions of CD4, CD38, CD56, CD123, and HLA-DR, but lack CD117. Further exploration of the immunophenotypic relationship may shed some light on the mechanism of their transformation.

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Blastic plasmacytoid dendritic cell neoplasm associated with chronic myelomonocytic leukemia

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