A CD34-negative MYC-rearranged B-lymphoblastic lymphoma aberrantly expresses CD3 and CD5

The patient is a 65-year-old man with HIV/AIDS. In February 2016, he presented to an outside hospital with ascites (panel A; hematoxylin and eosin stain, original magnification ×400) and a diagnosis of MYC-rearranged (panel B) CD5-dimly positive (panel C) “aggressive large B-cell lymphoma.” There was no CD3 (panel D) or rearrangement of BCL2 or BCL6. He was treated and his ascites resolved, but a follow-up scan many months later showed a renal lesion. A renal core biopsy showed medium-sized lymphoid blasts (panel E; hematoxylin and eosin stain, original magnification ×400), positive for CD10 (panel F; original magnification ×400), CD19 (panel G; original magnification ×400), terminal deoxynucleotidyl transferase (TdT; panel H; original magnification ×400), and cellular myelocytomatosis viral oncogene (panel I; original magnification ×400), with coexpression of CD5 (panel J; original magnification ×400) and nearly 100% Ki-67 (panel K; original magnification ×200), negative for CD34 (not shown). Interestingly, these blasts were dimly positive for CD3 (panel L; original magnification ×400). Polymerase chain reaction showed monoclonal IGK rearrangement (panel M, lanes 1 and 2) with germ line TRG; thus, a diagnosis of B-lymphoblastic lymphoma (B-LBL) was established. Retrospective immunohistochemistry of the ascites specimen pulled in February 2016 showed the blasts were positive for TdT (panel N; original magnification ×400).

This is an interesting case of B-LBL with several unusual features: in an HIV-positive patient, an initial presentation as ascites but later as a renal mass without bone marrow or blood involvement, and the first case of B-LBL with aberrant expression of CD5 and acquisition of CD3, the latter of which could be caused by lineage infidelity. Because of TdT immunoreactivity, this case shall not be diagnosed as high-grade B-cell lymphoma, not otherwise specified, as specifically defined by the 2016 revision of the World Health Organization’s classification of lymphoid neoplasms.
A CD34-negative MYC-rearranged B-lymphoblastic lymphoma aberrantly expresses CD3 and CD5

Huan-You Wang and John A. Thorson