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Occult ALK-negative anaplastic large cell lymphoma complicating chronic lymphocytic leukemia

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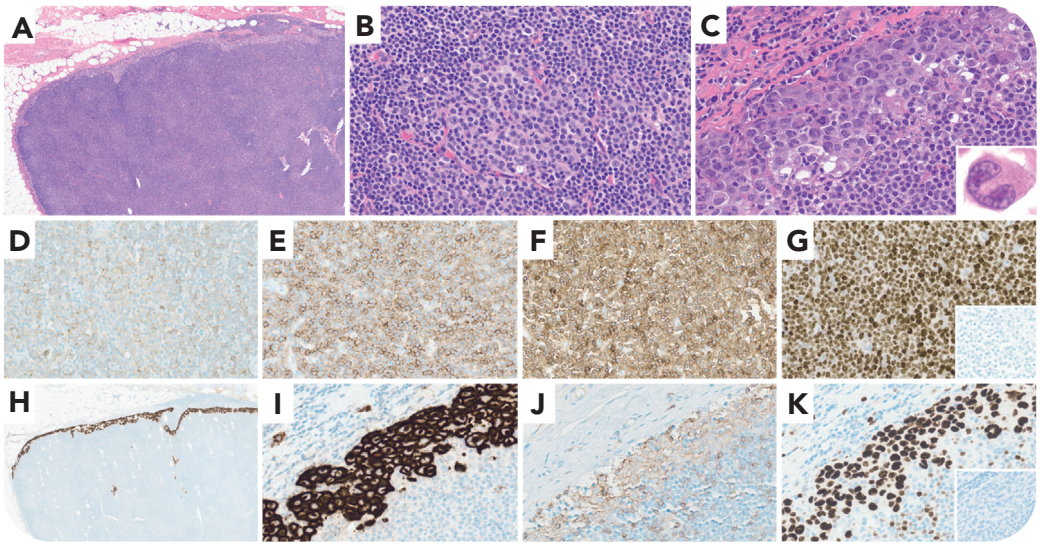

A 58-year-old man with a history of chronic lymphocytic leukemia (CLL) presented with 2 months of abdominal pain and night sweats. Imaging showed hypermetabolic abdominal, cervical, and axillary lymphadenopathy. Excisional biopsy of a 2.7-cm right axillary lymph node showed effacement of the lymph node architecture by a diffuse infiltrate of small lymphocytes with scattered proliferation centers (panel A, hemotoxylin and eosin [H&E] stain, 4× lens objective; panel B, H&E stain 40× lens objective). The sinusoids contained large atypical cells, including "hallmark" cells (panel C, H&E stain, 40× lens objective; inset, 100× lens objective). By immunohistochemistry, the small cells were positive for CD20 (weak; panel D, 40× lens objective), PAX5, CD5 (panel E, 40× lens objective), CD23 (panel F, 40× lens objective), and LEF1 (panel G, 40× lens objective) and negative for cyclin D1 (panel G, 40× lens objective; inset). The large cells were positive for CD30 (panel H, 4× lens objective; panel I, 40× lens objective) and weakly positive for CD2 and CD4 (panel J, 40× lens objective). Ki67 showed a high proliferation rate (panel K, 40× lens objective). They were negative for CD20, CD79a, PAX5, CD138, CD3, CD5, CD8, TIA1, granzyme B, and ALK (panel K, 40× lens objective; inset). Epstein-Barr virus encoded RNA was negative. Fluorescence in situ hybridization for *DUSP22* and *TP63* rearrangements was negative. These features indicated a composite lymphoma comprising CLL and ALK-negative anaplastic large cell lymphoma (ALCL).

Clinical progression in patients with CLL raises concern for transformation to aggressive B-cell lymphoma (Richter syndrome). De novo ALCL complicating CLL is uncommon but may occur. Some ALCLs present with a predominantly sinusoidal pattern, and pathological features may be subtle. Careful morphologic and immunophenotypic evaluation is essential for accurate diagnosis, which is critical because ALCL requires distinct therapy.

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