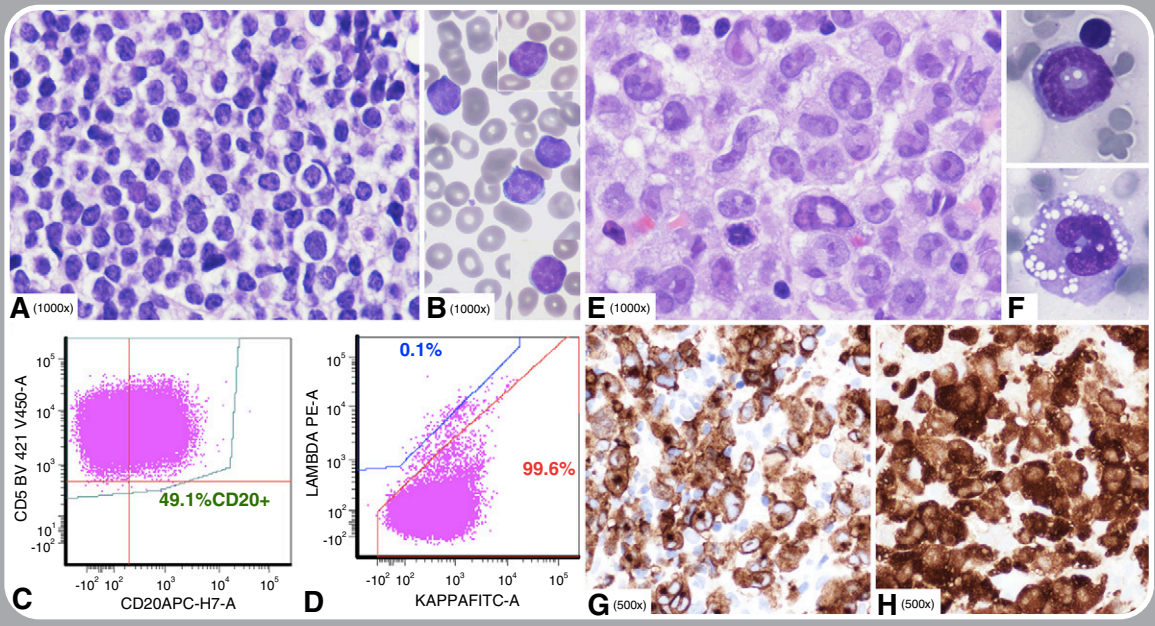
bloodwork
IMAGES IN HEMATOLOGY

"Soccer ball" cells to "donut" cells: an unusual case of Richter syndrome



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A 56-year-old man presented with relapsed chronic lymphocytic leukemia (CLL) with 90% bone marrow (BM) involvement, "soccer ball" morphology (panel A, hematoxylin and eosin stain; panel B, Wright-Giemsa stain), and the following immunophenotype: $CD5^+/CD19^+/CD20dim^+/CD38^+/ZAP\text{-}70^+$/dim κ (panel C [APC, allophycocyanin; BV, brilliant violet]; panel D [FITC, fluorescein isothiocyanate; PE, phycoerythrin]). Fluorescence in situ hybridization (FISH) detected deletion D13S319 in both loci. The *IGVH* gene was unmutated and next-generation sequencing (NGS) showed *SF3B1* mutation. He had a good response with venetoclax. Seven months later, he presented with pancytopenia, increased lactate dehydrogenase (1039 U/L), and multicompartmental lymphadenopathy. BM (panel E, hematoxylin and eosin stain) and subsequent lymph node biopsy showed large pleomorphic cells with "donut," horseshoe, and Reed-Sternberg Hodgkin–like morphology (panel F [original magnification, ×1000; Wright-Giemsa stain]; inset [original magnification, ×1000; Wright-Giemsa stain]) expressing CD30 (panel G, immunostain), ALK-1 (panel H, immunostain), and CD2 (weak)/CD4/CD5/CD45 (LCA)/CD43; he was negative for CD3, CD7/CD8/CD15, and Pax-5. Flow cytometry detected a minute residual CLL population (0.2%). FISH detected *ALK* rearrangement. The FISH panel demonstrated extra copies of *TP53* and *D12Z3* and monosomy 13. NGS detected 2 *TP53* gene mutations (TP53p.V216L and TP53p.L289fs) not identified previously.

Development of an aggressive neoplasm occurs in ~5% of patients with CLL and is known as Richter syndrome, with diffuse large B-cell lymphoma being the most common subtype. In 80% to 90% of cases, aggressive neoplasm is clonally related to CLL; in 10% to 20%, the clones are unrelated. This case illustrates anaplastic large cell lymphoma, ALK^+ as a rare subtype of Richter syndrome; ancillary studies suggest that the neoplasms are clonally unrelated.

DOI 10.1182/blood-2017-08-802900