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A peculiar case of cyclin D1–positive lymphoplasmacytic lymphoma

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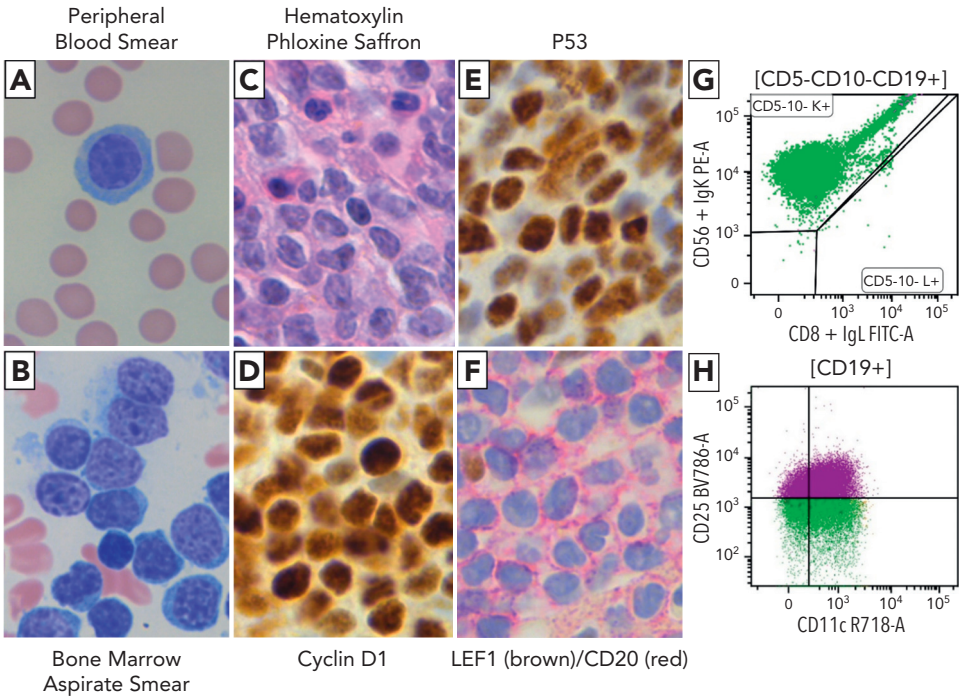

A 76-year-old man previously diagnosed with lymphoplasmacytic lymphoma (LPL) upon lymph node biopsy alongside immunoglobulin M (IgM) paraproteinemia and hyperviscosity was initially treated with rituximab, cyclophosphamide, vincristine, and prednisone followed by rituximab-bendamustine at relapse. The LPL was negative for cyclin D1 during the initial diagnosis. He presented again with new omental and mesenteric masses, pancytopenia, lymphocytosis (panel A, 63× objective), serum IgM-κ M-protein, and a faint IgA-κ band. Bone marrow aspirate and biopsy were hypercellular, showing 90% interstitial and paratrabecular involvement by κ-restricted medium-sized B cells with plasmacytoid differentiation (panels B-C, 63× objective; panel G), and 1% polytypic background plasma cells. The lymphoma strongly expressed cyclin D1 and p53 (panel D-E, 63× objective) and was negative for CD5/CD10/SOX11/LEF1/CD23 (panel F, 63× objective; panel G) with partial/dim CD25/CD103/CD11c (panel H). Fluorescence in situ hybridization study was negative for *CCND1*::*IGH* rearrangement. Therefore, small lymphocytic lymphoma, hairy cell leukemia, and mantle cell lymphoma were excluded. The strong expression of CD19/CD20/CD79a/CD45 and lack of CD138/CD38 excluded lymphoplasmacytic variant of plasma cell myeloma. Next generation sequencing confirmed a pathogenic variant of *TP53* (p.Val173Met) but no *MYD88* hotspot mutation.

The findings were compatible with recurrent *MYD88*^{WT} LPL with acquired cyclin D1 expression and *TP53* mutation. The patient responded well to zanubrutinib with significant shrinkage of masses and reduction of serum IgM. To our knowledge, this represents the first reported instance of cyclin D1-expressing LPL in the literature.

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