blood work
images in hematology

Primary thymic marginal zone lymphoma with amyloidosis

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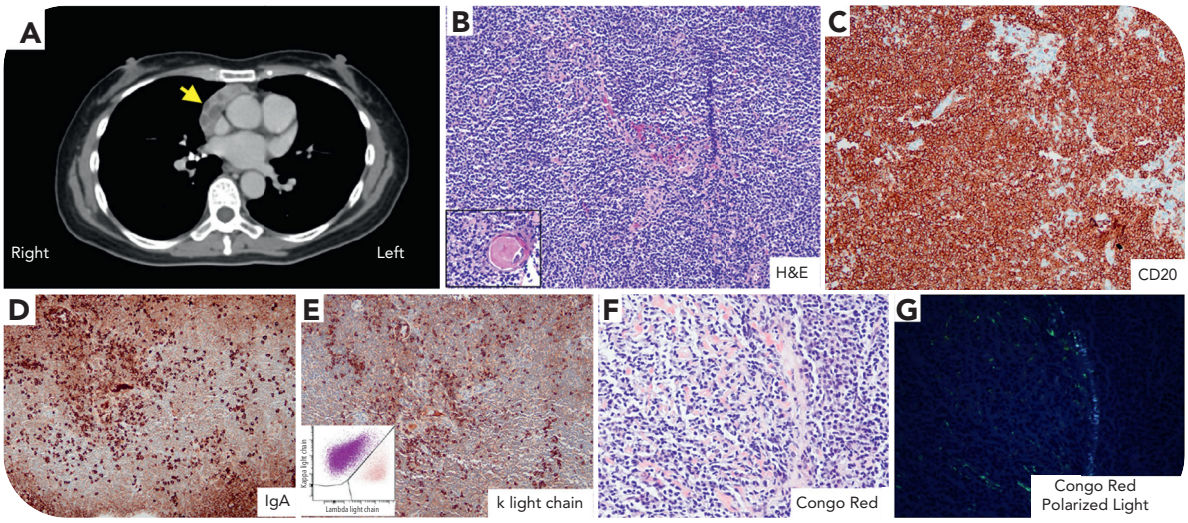

A 57-year-old woman with a history of Sjögren syndrome had an incidental 5.4 × 2.4-cm anterior mediastinal mass on computed tomography (panel A, arrow). A core biopsy demonstrated residual thymic tissue infiltrated by small to medium-sized atypical lymphoid cells with condensed chromatin, inconspicuous nucleoli, and scant cytoplasm (panel B and inset showing Hassall corpuscles, hematoxylin and eosin stain; 20× objective, ×200 total magnification). The neoplastic cells were positive for CD20, immunoglobulin A (IgA) heavy chain, and κ light chain (panels C-E; 20× objective, ×200 total magnification) and negative for IgM, IgG, and λ light chain (data not shown). Flow cytometry confirmed the presence of a κ-restricted B-cell population (panel E, inset), which was negative for CD10 and CD5 (data not shown). Congo red special stain was positive for amyloidosis (panels F-G; 40× objective, ×400 total magnification). Fluorescence in situ hybridization showed no evidence of *MALT1* rearrangement. Molecular studies revealed mutation in *CARD11*, exon 6 p.L251P(c.752T>C). Peripheral blood, bone marrow, and lymph nodes were uninvolved. Laboratory evaluation showed elevated IgA (1501 mg/dL), IgG (3044 mg/dL), and ratio of serum free κ (4.77 mg/dL) to λ (2.03 mg/dL) light chain (2.35). Protein electrophoresis and immunofixation detected IgG κ/IgA κ biclonal gammopathy. There was no clinical suspicion of systemic amyloidosis.

Primary thymic marginal zone lymphoma with amyloidosis is rare but nearly exclusively associated with Sjögren syndrome and has favorable prognosis.

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