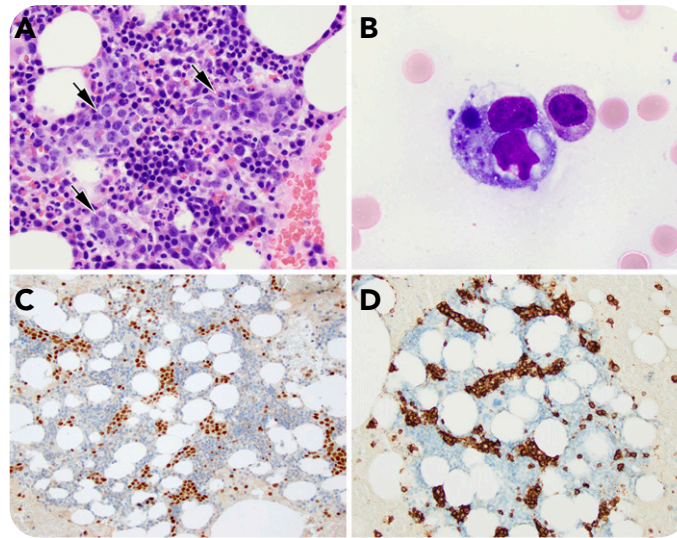


Hemophagocytic syndrome–associated intravascular large B-cell lymphoma

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The patient is a 66-year-old Middle Eastern woman who presented with B symptoms (severe fatigue, night sweats, and fever), weight loss, anemia, and splenomegaly with elevated liver enzymes and respiratory insufficiency. Bone marrow biopsy showed a B-lymphocyte population with pleomorphic nuclei and prominent nucleoli that expanded the bone marrow sinusoids (panel A, hematoxylin and eosin stain, 40× objective, original magnification ×400) and was highlighted by PAX5 (panel C, PAX5 immunohistochemical study, 20× objective, original magnification ×200) and CD20 (panel D, CD20 immunohistochemical study, 20× objective, original magnification ×200). Hemophagocytosis was present (panel B, Wright-Giemsa stain, 60× objective, original magnification ×600). Immunohistochemical studies showed the lymphoma cells to be positive for CD20, PAX5, CD5, BCL2, BCL6 (dim), MUM1, and LEF1 and negative for CD10, SOX11, c-MYC, and PD-L1. No other tissue biopsy was performed.

Intravascular large B-cell lymphoma is a rare aggressive lymphoma that shows growth within small vessels often with widespread dissemination. Three clinically distinct variants have been identified: classical variant (with frequent central nervous system, cutaneous, and endocrine involvement), cutaneous variant (with involvement limited to the skin), and hemophagocytic syndrome (HPS)-associated variant (with a typical HPS presentation). Previously, the HPS-associated variant was described as the Asian variant due to its near-exclusive presentation in patients of Asian ancestry, as in this case. Despite the different extent of involvement between the classical and HPS-associated variants, prognosis in both is extremely poor with this aggressive lymphoma. Recent studies have shown frequent mutations in *CD79B* and *MYD88*, which may present future avenues for targeted therapy.