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Hemophagocytic lymphohistiocytosis with multiple possible etiologies including rare primary bone marrow lymphoma

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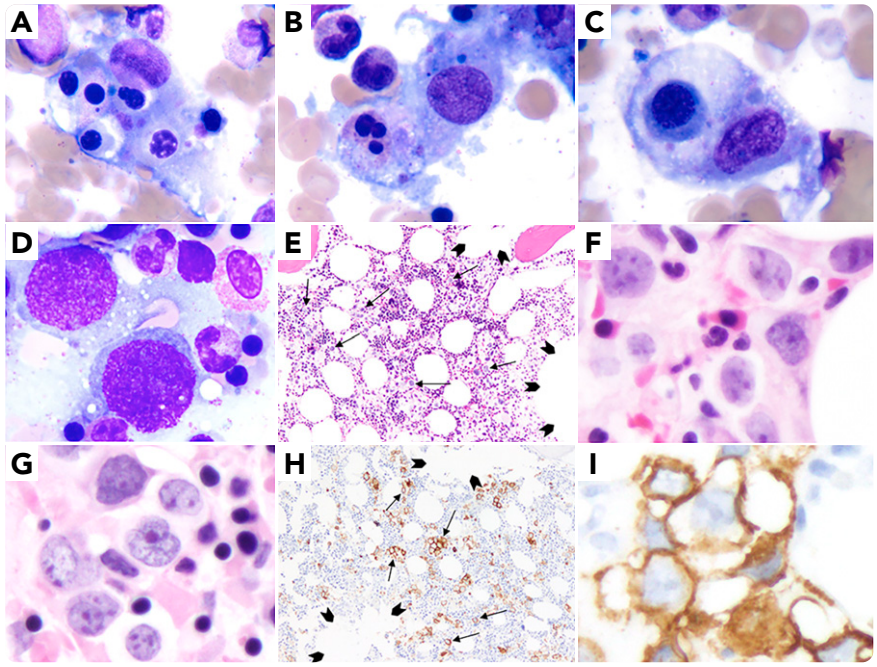

A 67-year-old woman presented with fatigue, fever (38.6°C), bicytopenia (platelets 9 × 10^3/μL; hemoglobin 9.6 g/dL), elevated ferritin (4313 ng/mL)/triglycerides (481 mg/dL)/interleukin-2 receptor (CD25: 87 950.0 pg/mL), cytomegalovirus viremia, and positive nasal swab for methicillin-resistant *Staphylococcus aureus*. A computed tomography scan showed hepatosplenomegaly without lymphadenopathy. Bone marrow aspirate smears showed hemophagocytic histiocytes (panels A-C: May-Grunwald-Giemsa, 100× objective, ×1000 magnification), large cells (panel D: May-Grunwald-Giemsa, 100× objective, ×1000 magnification), and negative flow cytometry. Trephine biopsy showed large cell infiltrate (panel E: hematoxylin and eosin (H&E), ×10X objective, 100× magnification; and panels F, G: H&E, 40× objective, ×400 magnification) with CD20 expression showing interstitial (not intravascular/intrasinusoidal) infiltration by lymphoma cells (arrows) and uninvolved sinusoids (arrowheads) (panel H: CD20, 10× objective, ×100 magnification showing interstitial distribution of large B cells; and panel I: CD20, 40× objective, ×400 magnification)/Bcl-2/Bcl-6/MUM1/MYC (not shown) and negative for CD10/CD5/cyclin-D1/CD30/ALK-1/TdT/CD34. Primary bone marrow diffuse large B-cell lymphoma (nongerminal center B cell) with hemophagocytic lymphohistiocytosis (HLH) was diagnosed (splenomegaly/fever/elevated ferritin/hypertriglyceridemia/elevated CD25/hemophagocytosis on aspirate met 6 of 8 criteria). Cytogenetics showed complex karyotype. After chemotherapy and antimicrobial coverage for methicillin-resistant *S aureus* and cytomegalovirus infections, the patient is doing well.

The case illustrates the importance of having a very high index of suspicion for malignancy in older patients with HLH even if there are other possible nonmalignant (eg, infections) causes of HLH for appropriate therapy.

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