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Immune dysregulation: EBV$^+$ DLBCL and HLH in a patient with T-LGL

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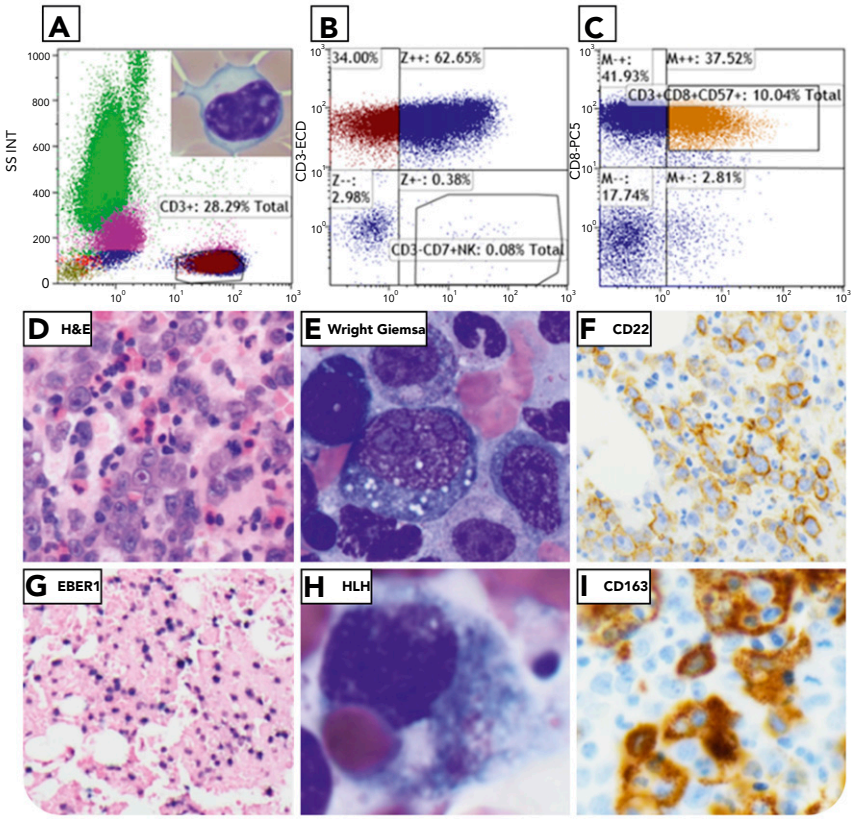

An 80-year-old man with a 20-year history of T-cell large granular lymphocytic leukemia (T-LGL), receiving low-dose neupogen for neutropenia, presented with new-onset fever, pancytopenia, and tachycardia. Flow cytometry of the peripheral blood showed aberrant clonal T cells (6% to 8% of total lymphocytes; panels A-C), CD3$^+$, CD8$^+$, CD7$^-$, CD4$^-$, CD57$^+$ (panel A inset LGL, 100× objective), supporting T-LGL. Bone marrow (BM) biopsy (panel D, 40× objective) and smear (panel E, 100× objective) showed large pleomorphic cells with prominent nucleoli and cytoplasmic vacuoles CD22$^+$ (panel F), PAX5$^+$, CD79A$^+$, MUM1$^+$, CD30$^+$, and CD10$^-$ (not shown). T-LGL cells were persistent in the BM, CD3$^+$, CD8$^+$, and CD57$^+$. EBER1 in situ hybridization–positive cells (panel G) supported Epstein-Barr virus (EBV) diffuse large B-cell lymphoma (EBV$^+$ DLBCL), non–germinal center type. Interphase fluorescence in situ hybridization was negative for BCL2, BCL6, and MYC rearrangements. Hemophagocytosis (HLH) was seen on BM smear (panel H, 100× objective), highlighted by CD163 (panel I) on biopsy. Ferritin was 27 519 ng/mL (reference, 30-400 ng/mL); aspartate aminotransferase, 59 U/L (reference, 1-35 U/L); lactate dehydrogenase, 1041 U/L (reference, 100-200 U/L); and fibrinogen, 99 mg/dL (reference, 175-450 mg/dL), supporting HLH.

T-LGL is a rare chronic clonal mature T-cell neoplasm, commonly coexisting with clonal B-cell processes. In this patient, EBV$^+$ DLBCL most likely caused secondary HLH, an ultimate cytokine storm, and immune dysregulation. The patient died soon after.

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