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Plasma cell leukemia with small cell morphology

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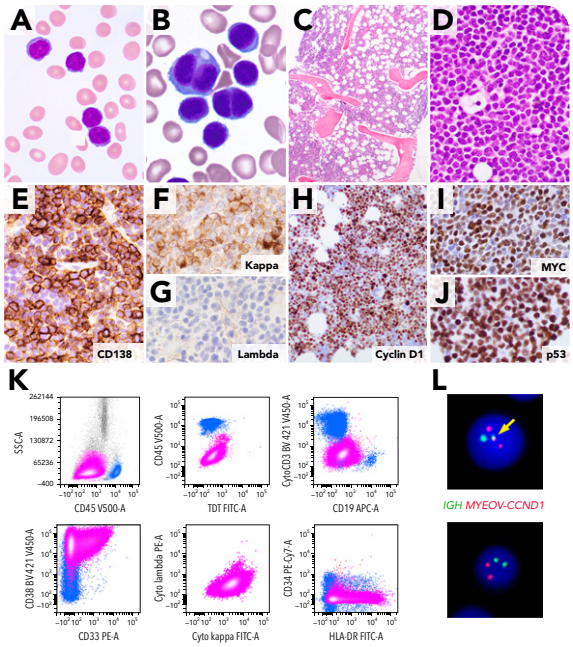

A 64-year-old woman with a history of Hodgkin lymphoma treated 2.5 years ago presented with worsening leukocytosis (60 700-147 000/μL) over 3 months but without lymphadenopathy, lytic bone, or hypermetabolic lesions (presenting hemoglobin, 6.6 g/dL; platelets, 24 000/μL). Serology showed no M-spike, but it did show elevated free kappa (64.88 mg/L) and an increased free kappa/lambda ratio (24.86). Urine studies confirmed kappa Bence-Jones proteinuria (32 mg/day). A peripheral blood smear showed many small lymphoid-appearing cells (97%), some with binucleation or perinuclear clearing (panel A, ×1000 original magnification). Bone marrow aspirates revealed an increase in similar cells to 72% (panel B, ×1000 original magnification); biopsy showed sheets of plasma cells (PCs, 70%-80%) (panel C, ×40 original magnification; panel D, ×500 original magnification), expressing CD138/cyclin D1/MYC/P53/kappa and not expressing CD5/CD20/immunoglobulin M [IgM]/IgA/IgD lambda (E-G, I-J, ×500 original magnification; H, ×200 original magnification). Flow cytometry confirmed cytoplasmic kappa-restricted PCs expressing CD33/CD38/CD81(partial)/CD138/HLA-DR and not expressing CD19/CD27/CD34/CD45/CD56/CD117/terminal deoxynucleotidyl transferase (TdT) (K, pink population). Fluorescence in situ hybridization identified *IGH-MYEOV/CCND1* fusion (L, arrow), deletions of *CDKN2C/RB1/MAF/TP53* and 3′ *MYC*, and monosomies 1 and 17. Chromosome analysis showed a hypodiploid complex karyotype. Next-generation sequencing detected *TP53* mutation (p.C238G). Plasma cell leukemia (PCL) was diagnosed and treated with carfilzomib-hypercytoxan-dexamethasone. At 1-month follow-up, leukocytosis improved (13 200/uL) but with persistent circulating PCs (51%). She died 1 month later.

PCL presenting with hyperleukocytosis and small cell morphology without extramedullary involvement is unusual and can be diagnostically challenging by mimicking acute lymphoblastic leukemia and leukemic non-nodal mantle cell lymphoma.

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