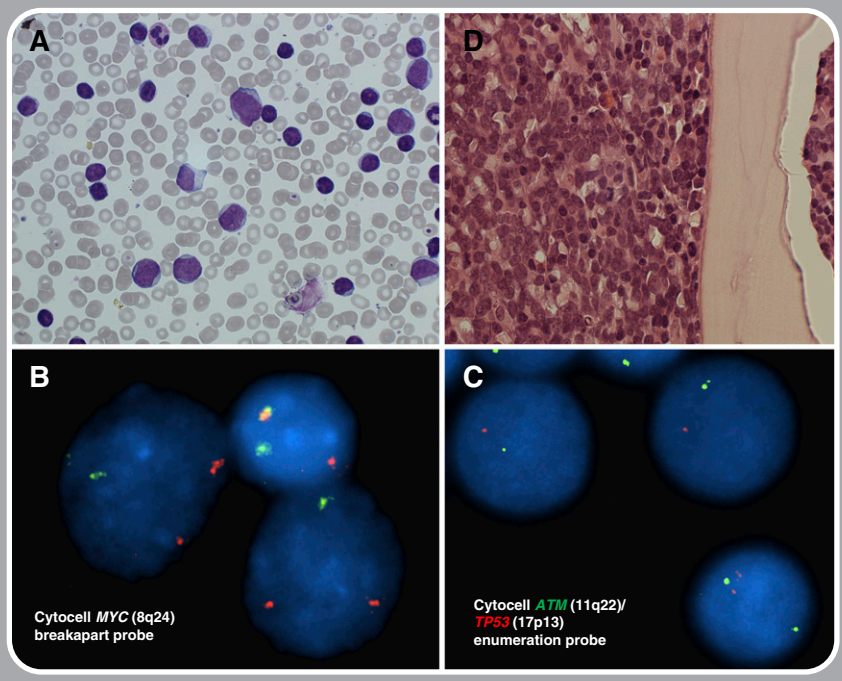
bloodwork
IMAGES IN HEMATOLOGY

De novo Richter transformation



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A 68-year-old woman presented with pneumonia and acute kidney injury. White blood cell count was 133.4 × 10^9/L (lymphocytes, 92.5 × 10^9/L). Blood count a month previously had shown mild lymphocytosis (9.6 × 10^9/L). Peripheral blood film showed 2 cell populations of small lymphocytes and 20% larger blast-like cells with small nucleoli (panel A; original magnification ×50, May-Grünwald-Giemsa stain) and morphology distinct from prolymphocytes, which typically have 1 large nucleolus. Peripheral blood immunophenotyping revealed 2 surface immunoglobulin κ restricted clonal B-cell populations, both positive for CD19/CD23/CD20/CD22/CD79b/CD38; the population with low forward scattered light was $CD5^+$/FMC^-; the other was $CD5^-$/$FMC7^+$. *IGHV-D-J* rearrangement analysis confirmed the 2 populations were clonally related. Peripheral blood and bone marrow fluorescence in situ hybridization detected abnormalities in 90% cells: smaller cells (70%) had standard *MYC* (8q24) rearrangement without *ATM* (11q22) or *TP53* (17p13) abnormalities, whereas larger cells (~20%) had both copies of *MYC* rearranged and additional *ATM* and *TP53* loss (panels B-C), consistent with aggressive B-cell malignancy arising through clonal evolution of chronic lymphocytic leukemia (CLL) or Richter syndrome (RS). Bone marrow biopsy (panel D; original magnification ×20, hematoxylin and eosin stain) showed aggressive B-cell lymphoma involvement.

RS occurs in 2% to 10% of CLL cases: determining whether RS is clonally related (80% cases) to CLL is critical, as these cases carry poorer prognosis. Forty percent of RS cases develop before requiring CLL treatment and may have somewhat improved outcomes.

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