

Images in Clinical Hematology

A rare type of acute leukemia in peripheral blood smear



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A 80-years-old woman presented with 6-weeks history of fatigue and shortness of breath. Hemoglobin was 9.7 g/dL, platelets were $79 \times 10^3 /\mu\text{L}$, leukocytes were $4.06 \times 10^3 /\mu\text{L}$ and B12 vitamin level was $<200 \text{ pg/ml}$ (reference value 197 - 771). After administration of cyanocobalamin her B12 vitamin level improved, but the anemia got progressively worse (hemoglobin was 3.7 g/dl) and reticulocyte count did not improve. A peripheral blood smear showed proerythroblasts (Figures 1, 2). Bone marrow aspirates and biopsy showed hypercellularity, being mainly erythroid progenitors, constituting $>80\%$ of bone marrow cell count with $>30\%$ proerythroblast without a significant myeloblastic component. The erythroid progenitors exhibited CD45+, CD34-, CD71+ and CD117+ by flow cytometry, and E-cadherin+, CD71+ and TP53+ by immunohistochemical staining. Fluorescence in situ hybridization analysis showed deletion 5q and loss of TP53. Karyotype showed deletion 5q, and derivatives chromosomes 17 (included loss of TP53 gen) and 19. Next-generation sequencing panel detected potentially pathogenic variants in TET2 (VAF 28%) and TP53 (VAF 40%) genes. The findings

were diagnostic of pure erythroblastic leukemia (PEL). Our patient was treated with azacytidine and blood transfusion support with a poor response over five months of follow-up.

PEL is a rare type of acute leukemia that represents less than 1% of all cases of acute myeloid leukemia (previously

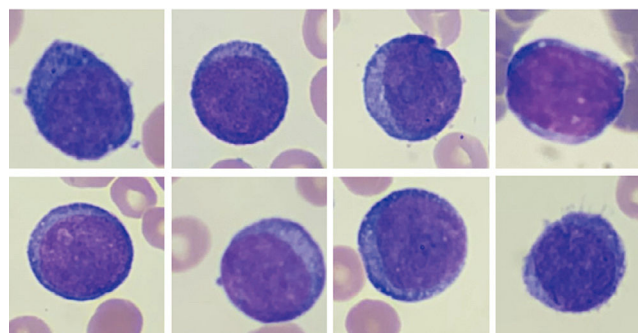


Figure 1 – Peripheral blood smear. Proerythroblasts; panel A-H: erythroid progenitors having large irregular nuclei, dispersed chromatin, some with prominent nucleoli, deeply basophilic cytoplasm, and high nuclear to cytoplasmic ratios. Wright stain; 100X objective, original magnification X1000.

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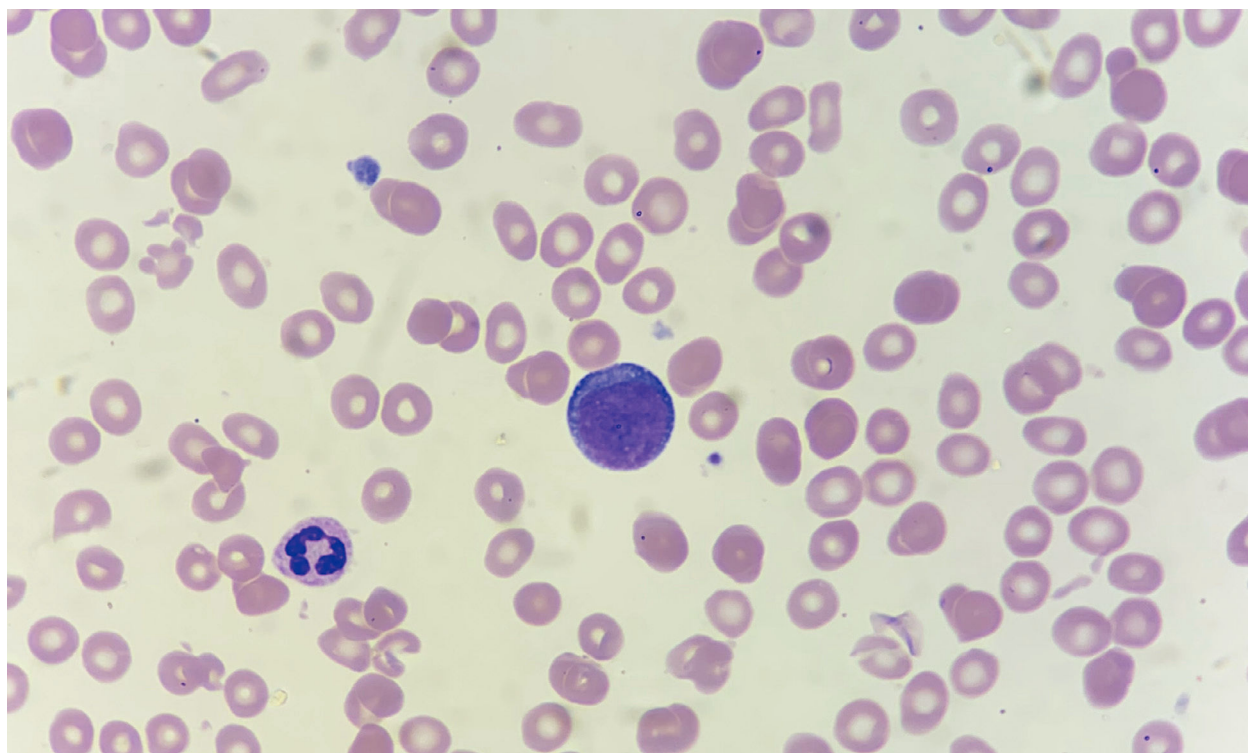


Figure 2 – Proerythroblast in peripheral blood smear. Wright stain; 100X objective, original magnification X1000.

called M6, by the French-American-British cooperative group).^{1,2} PEL may be therapy-related, preceded by a myelodysplastic syndrome or develop de novo.² PEL is defined in the 2016 WHO classification system, as a neoplastic proliferation of erythroid progenitors constituting > 80% of bone marrow cellularity with $\geq 30\%$ proerythroblasts without a significant myeloblastic component.³ Reactive erythroid hyperplasia is a well-known morphologic mimic of PEL in many diverse clinical situations due to erythroid hyperplasia as non-neoplastic (eg, megaloblastic anemia) and neoplastic entities. Although clinical presentation, laboratory, cytogenetic and molecular studies may ultimately resolve the differential diagnosis (eg, P53 mutation, complex karyotype).⁴ PEL have a clinically aggressive course associated with a poor prognosis.¹

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

Conflicts of interest

The authors declare no conflict of interest.

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