

## CLINICAL IMAGE

# Pure erythroid leukemia

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**Abstract**

Pure erythroid leukemia is a rare and aggressive form of acute leukemia with a deleterious clinical course. It is of erythroid lineage without myeloblastic component, representing >80% of marrow cellularity, with  $\geq 30\%$  proerythroblasts.

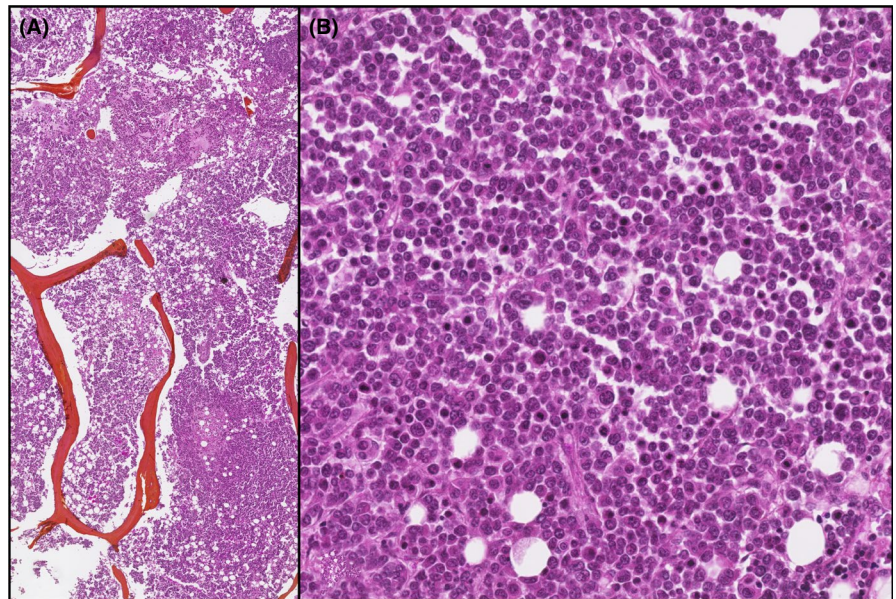
**KEYWORDS**

AML, bone marrow pathology, hematological oncology, pancytopenia, pure erythroid leukemia

A 60-year-old woman presented with lower gastrointestinal bleeding to the emergency department. She was a smoker, known for Crohn's disease and antiphospholipid syndrome treated with warfarin and mercaptopurine. Upon hospitalization, pancytopenia was discovered (leukocytes  $1.3 \times 10^9/L$ , neutrophils  $0.72 \times 10^9/L$ , hemoglobin  $64 \times 10^9/L$ , reticulocytes  $4.4 \times 10^9/L$ , and platelets  $57 \times 10^9/L$ ). Peripheral smear

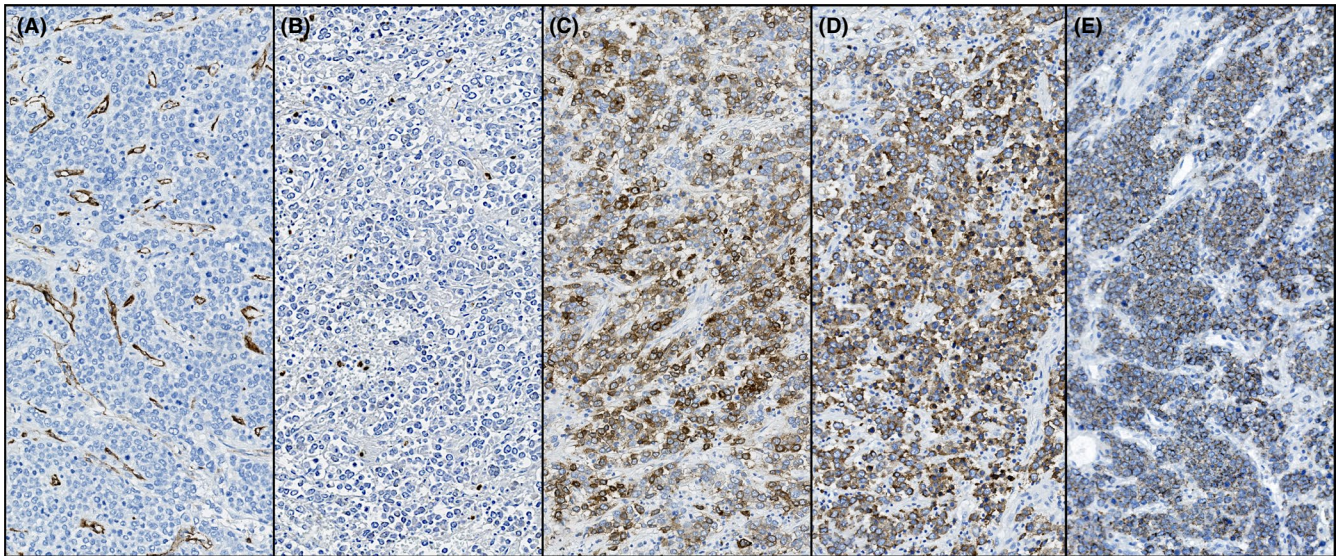
was interpreted as reactive changes with no myelodysplastic syndrome. Infectious and occult bleeding work-ups were negative. A bone marrow biopsy was deemed unsafe as patient was febrile and had desaturation episodes. Thoracic CT scan showed mild bilateral pleural effusion, without pneumonia or lymphadenopathy. She died 10 days after hospitalization from respiratory insufficiency and severe lactic acidosis.

**FIGURE 1** Bone marrow shows a markedly hypercellular marrow (90%) (A, HE,  $\times 20$  objective), infiltrated by numerous monomorphous neoplastic cells of erythroid lineage (B, HE,  $\times 200$  objective)



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**FIGURE 2** The neoplastic erythroblasts in the spleen were negative for CD34 (A, CD34,  $\times 200$  objective) and myeloperoxidase (B, myeloperoxidase,  $\times 200$  objective) and positive for CD117 (C, CD117,  $\times 200$  objective), CD71 (D, CD71,  $\times 200$  objective), and E-cadherin (E, E-cadherin,  $\times 200$  objective)

Autopsy findings included splenomegaly (484 g), hemorrhagic foci in lungs, heart, and gastrointestinal tract, and severe pulmonary edema and emphysema. Microscopic examination revealed infiltration of lungs, spleen, kidneys, liver, and lymph nodes by undifferentiated neoplastic cells. The bone marrow was hypercellular (90%) with  $>80\%$  neoplastic erythroid precursors (Figure 1). Neoplastic cells expressed CD117, CD71, and E-cadherin, but not CD45, CD34, MPO, TdT, pancyokeratin, and S100 (Figure 2). Diffuse TP53 immunohistochemical staining was demonstrated. A diagnosis of pure erythroid leukemia was made.

Pure erythroid leukemia (PEL) is a rare and aggressive form of acute leukemia with deleterious clinical course and median survival of  $<3$  months.<sup>1</sup> PEL is of erythroid lineage without myeloblastic component, representing  $>80\%$  of marrow cellularity, with  $\geq 30\%$  proerythroblasts.<sup>2</sup> It remains a challenging diagnosis that requires correlating clinical, pathological, and cytogenetic results.

#### ACKNOWLEDGMENTS

None. Written consent was obtained from next of kin, following institutional guidelines.

#### CONFLICT OF INTEREST

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication

of this article. Informed consent was obtained for this publication.

#### AUTHOR CONTRIBUTIONS

MF: collected data and wrote the manuscript. SFR: wrote the manuscript. MH and AM: supervised the study and reviewed the manuscript.

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