

# Acute Pure Erythroid Leukemia with Fulminant Hemophagocytosis: A Case Report and Literature Review

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## BACKGROUND

Acute erythroid leukemia (AML-M6) is an uncommon type of acute leukemia comprising 2- 5% of all acute leukemia cases. Two subtypes of acute erythroid leukemia exist: the mixed type (erythroid/myeloid) or AML-M6a, and pure erythroid leukemia or AML-M6b. The latter diagnosis is made when  $\geq 80\%$  of the nucleated cells in the bone marrow is of erythroid lineage and no evidence of a myeloblastic component. Hemophagocytosis has no known association with acute myeloid leukemias. There are only two other known reported cases of AML-M6 with active hemophagocytosis. The first case was a pediatric patient with AML-M6a. The second was an adult patient with de-novo AML-M6b. This patient is the first report of complications with active hemophagocytosis in pure erythroid leukemia arising from myelodysplastic syndrome.

## PATIENT HISTORY

We report a case of a 75 year old African American female who presented to our hospital with generalized weakness, easy bruising, weight loss, and fevers. She was admitted with anemia, thrombocytopenia, and leukocytosis with blasts in the peripheral blood. She had previously been diagnosed with myelodysplastic syndrome 4 years prior at another institution. She had received hydroxyurea and had a follow-up bone marrow 2 year prior with essentially the same findings. Both of these two bone marrows showed only inversion of chromosome 9. A bone marrow biopsy was performed at our institution for assessment of her bone marrow status.

## RESULTS

The bone marrow biopsy showed a hypercellular marrow with fulminant hemophagocytosis and increased blasts. By morphology and flow cytometry the blasts were identified as erythroid lineage. They are positive for glycophorin-A and negative for CD13, CD33, and Myeloperoxidase. She was diagnosed with acute pure erythroid leukemia (AML M6b).

## BONE MARROW ASPIRATE

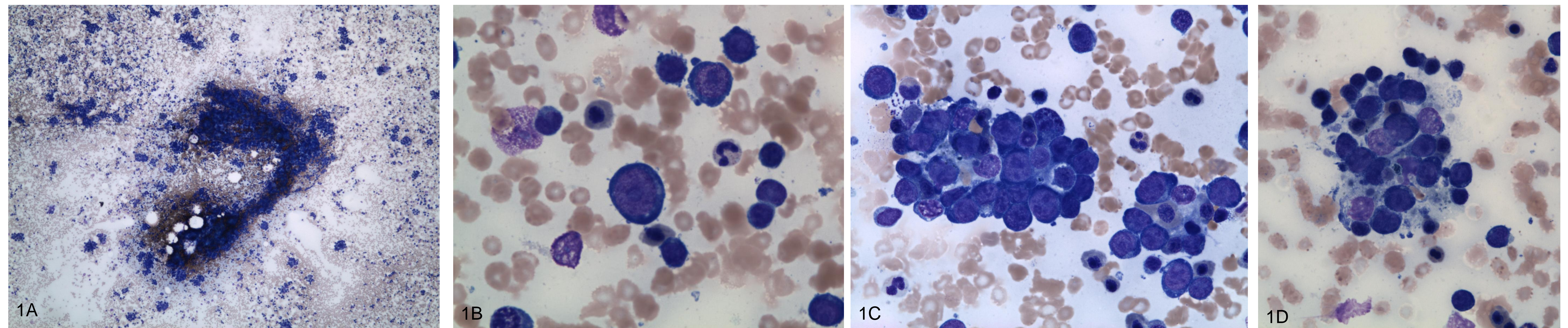


Fig 1A-D: (1A) Spicule with small groups of marrow elements (1B) Erythroid blast and precursors (1C-1D) Hemophagocytosis of erythroid blasts and precursors

## BONE MARROW CORE AND CLOT SECTIONS with FLOW CYTOMETRY

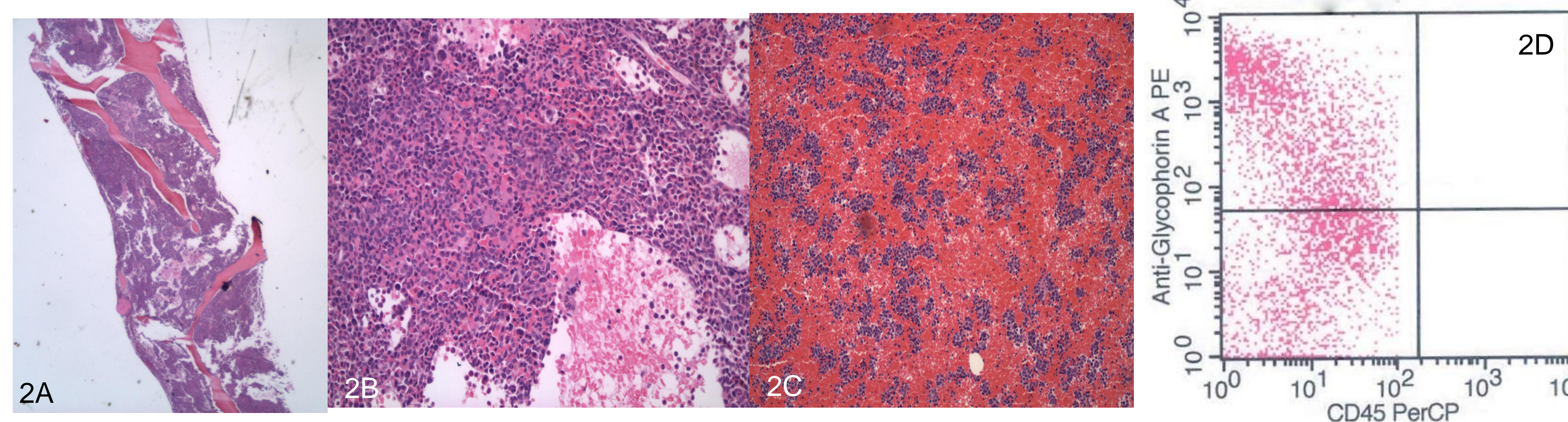


Fig 2A-D: Hypercellular bone marrow core and clot with monotonous cell population (2A core at 10x; 2B core at 20x; 2C clot at 10 x). 2D Flow cytometry scattergram with showing glycophorin –A positivity in the majority of the cells.

## CYTOGENETICS

Her chromosome studies revealed inversion of chromosome 9 along with the following cytogenetics: trisomy of chromosomes 1, 2, 6, 8, 13, and 21 and tetrasomy of chromosome 3. One copy of chromosome 3 had a deletion of the distal half of its long arm. One copy of chromosome 8 had additional chromatin of unknown origin on its short arm. A derivative chromosome was composed of the long arms of chromosomes 15 and 17 (the p53 gene locus).

## CONCLUSIONS

Acute pure erythroid leukemia is an uncommon subtype of acute leukemia. There have been only two other cases of acute erythroid leukemia with hemophagocytosis. This is the first case of AML-M6b with hemophagocytosis arising in the background of myelodysplastic syndrome. Hemophagocytosis can be an important cause of fevers in acute leukemic patients, although rare.

## References

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