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Peripheral Hemophagocytosis and Leukemic Blasts from Urine in De Novo Pure Erythroid Leukemia

De Novo Saf Eritroid Lösemide Çevresel Hemofagositoz ve İdrarda Lösemik Blastlar

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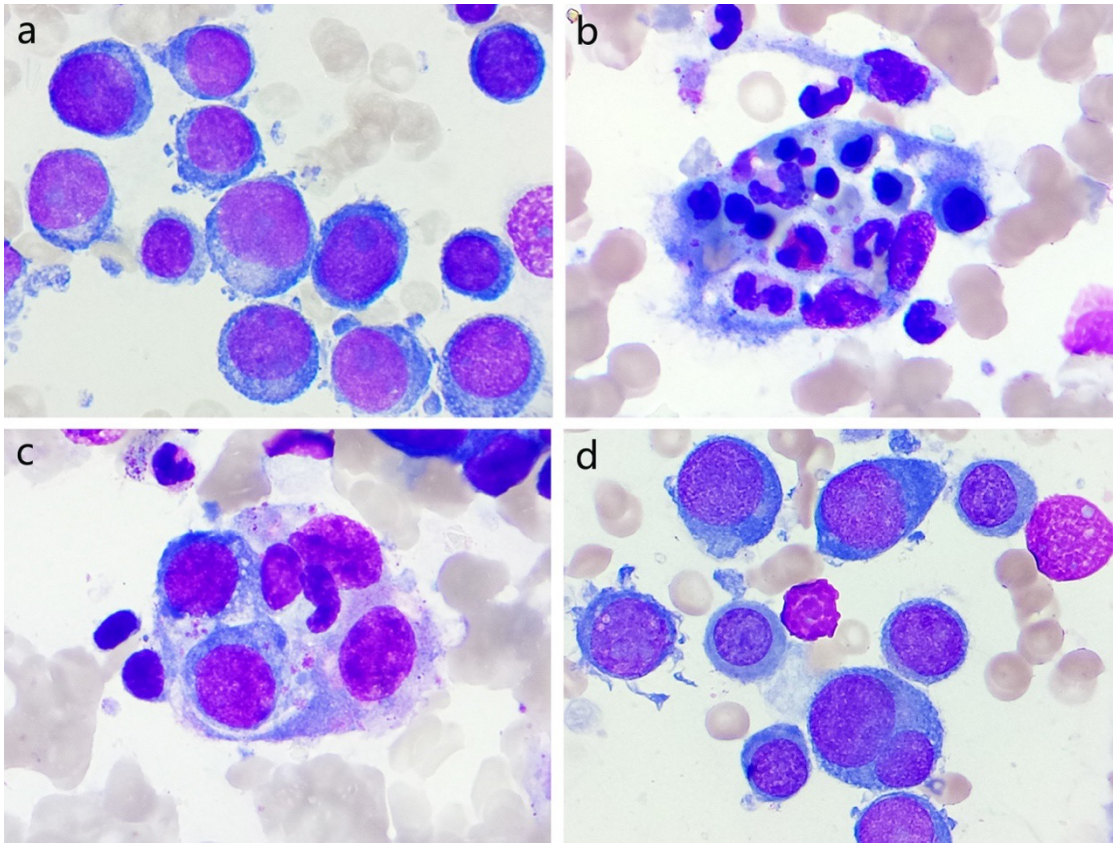

Figure 1. Peripheral blood smears revealed numerous blasts as well as hemophagocytes phagocytizing neutrophils, eosinophils, platelets, erythrocytes, and proerythroblasts (a, b, c, 1000^{x}, Wright-Giemsa staining). Urine smears showed leukemic blasts with morphological features consistent with the peripheral blood smears (d, 1000^{x}, Wright-Giemsa staining).

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A 46-year-old male patient with a history of hypertension for 4-5 years was admitted with complaints of bleeding gums and general fatigue for 7 days. A complete blood count showed white blood cell count of $47.95x10^9$/L, hemoglobin concentration of 55 g/L, and platelet count of $11x10^9$/L. Bone marrow aspiration showed 81% immature cells with the features of proerythroblasts. Immunophenotyping analysis demonstrated that the blast population was positive for CD36, CD105, CD33, CD117, CD13, CD71, CD34, HLA-DR, and CD38. Bone marrow biopsy indicated sheets of blasts and E-cadherin was positive by immunohistochemistry. Cytogenetics showed a complex karyotype with 52,XY,-4,-5,add(5)(q13),+14,add(15)(p11.2),-16,+19,add(19)(p13.1),add(19)(p13.1),-21,+mar1~8 [10]. Next-generation sequencing detected *TP53* mutation with a median variant allele frequency of 58%. A final diagnosis of pure erythroid leukemia (PEL) was made. The patient underwent chemotherapy with DA (daunorubicin, 60 mg, days 1-3; cytosine arabinoside, 0.1 g, every 12 h, days 1-7), and then cytarabine (38 mg, days 1-5) and another course of DA chemotherapy (daunorubicin, 60 mg, days 1-3; cytosine arabinoside, 0.1-5 g, once daily, days 1-7). However, he never achieved remission and rapidly progressed to multiorgan failure and macroscopic hematuria 77 days after the initial admission. Peripheral blood smears revealed numerous blasts as well as hemophagocytes phagocytizing neutrophils, eosinophils, platelets, erythrocytes, and proerythroblasts (Figures 1a-1c). Notably, urinalysis showed a cell count of 392.98/µL (reference range: 0-17/µL) and urine cytology was performed. Leukemic blasts were also noted in this urine specimen with morphological features consistent with the peripheral blood smears (Figure 1d). The results of urinalysis before the chemotherapy had been normal, with no leukocytes in the urine. Imaging of the kidneys and bladder showed no significant abnormalities, which ruled out the renal or bladder involvement of leukemia. Hence, the patient's hematuria was most likely due to severe thrombocytopenia. Additional laboratory tests to support hemophagocytic lymphohistiocytosis (HLH) were within normal reference values. Moreover, blood cultures of both aerobic and anaerobic bacteria were all negative several times throughout the whole clinical course, ruling out hemophagocytosis secondary to sepsis. Unfortunately, the patient died of myelosuppression, respiratory failure, liver and kidney dysfunction, gastrointestinal bleeding, and coagulation dysfunction within 3 months of the initial presentation.

The presence of peripheral hemophagocytosis, as an important laboratory indicator for the diagnosis of sepsis or secondary HLH, is quite rare in de novo PEL cases. As is well known, urothelial carcinoma is the most common tumor encountered in exfoliative urine cytology [1]. Leukemic invasion of the genitourinary system is a very rare event, but it may occur due to infiltration of the bladder with blast cells or as a result of hemorrhage in the urinary bladder caused by thrombocytopenia, with contamination of the urine by leukemic blasts attributable to an increased peripheral blast count [1,2]. Leukemic cells were found by accident in the routine urinalysis of this patient by microscopic examination. This case of the concurrence of peripheral hemophagocytosis and leukemic blasts in a urine specimen in de novo PEL was extremely uncommon. The presented case highlights the value of morphological methods providing simple, fast, and cost-effective approaches for the detection of peripheral hemophagocytosis and leukemic blasts in urine specimens.

Keywords: Peripheral hemophagocytosis, Leukemic blasts, Urine specimen, De novo pure erythroid leukemia

Anahtar Sözcükler: Çevresel hemofagositoz, Lösemik blastlar, İdrar örneği, De novo saf eritroid lösemi

Ethics

Informed Consent: Informed consent was obtained from this patient.

Footnotes

Authorship Contributions

Surgical and Medical Practices: M.Y., Y.T., F.H., L.Z., J.L., X.L.; Concept: F.H., L.Z., J.L., X.L.; Design: F.H., L.Z., J.L., X.L.; Data Collection or Processing: F.H., L.Z., J.L., X.L.; Analysis or Interpretation: M.Y., Y.T., F.H.; Literature Search: M.Y., Y.T., F.H.; Writing: M.Y., Y.T., F.H.

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