

Massive Intravascular Hemolysis and a Rapidly Fatal Outcome*

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A 58-year-old man 1 year status post allogeneic stem cell transplant for acute myeloid leukemia was admitted to the hospital for chemotherapy (cytosine arabinoside, etoposide, and mitoxantrone) due to a relapse. Except for fatigue, his review of symptoms was negative. His medications included prednisone and tacrolimus. Physical examination findings were unremarkable. He was comfortable, his mental status was intact, conjunctiva was not icteric, chest sounds were clear, there were no heart murmurs, abdomen was nontender, and he had no suspicious cutaneous lesions. A hemogram was significant for a platelet count of 26,000 cells/mL. On hospital day 5, he became neutropenic as a result of the chemotherapy with a WBC count that fell to 400 cells/mL. On day 7 of therapy, he began experiencing intense full-body rigors; hematuria and jaundice developed, followed by increasing oxygen requirements, sinus tachycardia (140 beats/min), a fever to 103°F, and hypotension (BP, 80/62 mm Hg). Arterial blood gas values with the patient breathing 60% O₂ revealed a pH of 7.56, a PaCO₂ of 29 mm Hg, and a PaO₂ of 155 mm Hg.

Several attempts to measure laboratory indices failed because of complete hemolysis of the patient's blood samples. His BUN and creatinine levels rose to 41 and 1.8 mg/dL, respectively (from 23 and 0.9

mg/dL, respectively), with concomitant hyperphosphatemia (7.4 mg/dL) and hypocalcemia (6.9 mg/dL). The hematocrit fell to 10.4% from 25.7% that morning. Levels of aspartate and alanine aminotransferases were also elevated over the short term from normal levels to 713 and 187 IU/L, respectively. The total bilirubin concentration increased to 8.4 mg/dL, the INR rose to 3.88, and the fibrin splits were > 40. Urine samples were bloody, and urinalysis revealed 3+ protein. Chest radiography demonstrated new bilateral perihilar infiltrates. The ECG showed lateral ischemia. He was transferred to the ICU, intubated, and given IV fluids, pressors, vancomycin, and piperacillin/tazobactam. The peripheral blood smear demonstrated marked spherocytosis (Fig 1) and the presence of Gram-positive bacilli (Fig 2). Clindamycin was added to the therapy. Aggressive transfusion was undertaken with multiple rounds of fresh-frozen plasma, cryoprecipitates, platelets, packed RBCs, and vitamin K without response. The patient required dual-pressor support.

What is the diagnosis?

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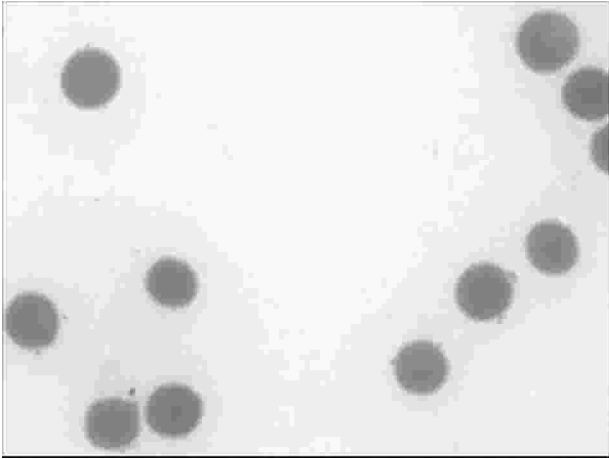


FIGURE 1. A predominance of spherocytes was noted on high-powered magnification of the peripheral blood smear (Wright-Giemsa stain, original $\times 1,000$).

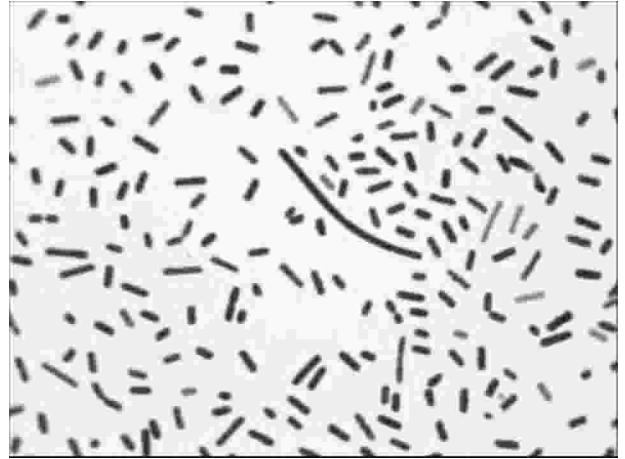


FIGURE 2. A colony from a blood culture (Gram stain, original $\times 500$).

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