An Unusual Case of Acute Hemolysis in a Jehovah's Witness After Stem Cell Transplant

A. A. Mitma[1], A. Venkata[1], L. F. Carrillo[2], F. van Rhee[3]; [1]Department of Internal Medicine, Division of Pulmonary and Critical Care Medicine, University of Arkansas for Medical Sciences, Little Rock, AR, United States, [2]Department of Pathology, University of Arkansas for Medical Sciences, Little Rock, AR, United States, [3]Department of Internal Medicine, Division of Hematology/Oncology, The Myeloma Center, University of Arkansas for Medical Sciences, Little Rock, AR, United States.

Introduction:Hemolysis remains an important differential diagnosis for patients admitted to ICU with severe anemia. In all cases of hemolysis, drug-induced hemolysis needs to be evaluated as a potential cause. Case Description: A 56-year-old female who is Jehovah's witness, with past medical history of lupus, received an autologous stem cell transplant for multiple myeloma after conditioning with high dose melphalan. The uric acid was noted to be trending up to 7.9 mg/dL and she received 3 mg of intravenous rasburicase on day 5 post-transplant. Subsequently, the patient was noted to have orange-colored urine and dark urine the next day. This was associated with an acute drop in hemoglobin from 11 to 7 g/dl. Urinalysis showed large amount of blood but normal RBCs. There was no other source of bleeding identified. She was transferred to the ICU due to severe anemia in the setting of recent myeloablative chemotherapy. She was started on intravenous steroids for possible autoimmune etiology. Workup revealed very low haptoglobin (6 mg/dL) and elevated indirect bilirubin (1.9 mg/dL). The direct coombs came back negative. Peripheral smear showed numerous eccentrocytes, no schistocytes, and occasional spherocytes; morphologic findings consistent with oxidant hemolysis (Figure A).

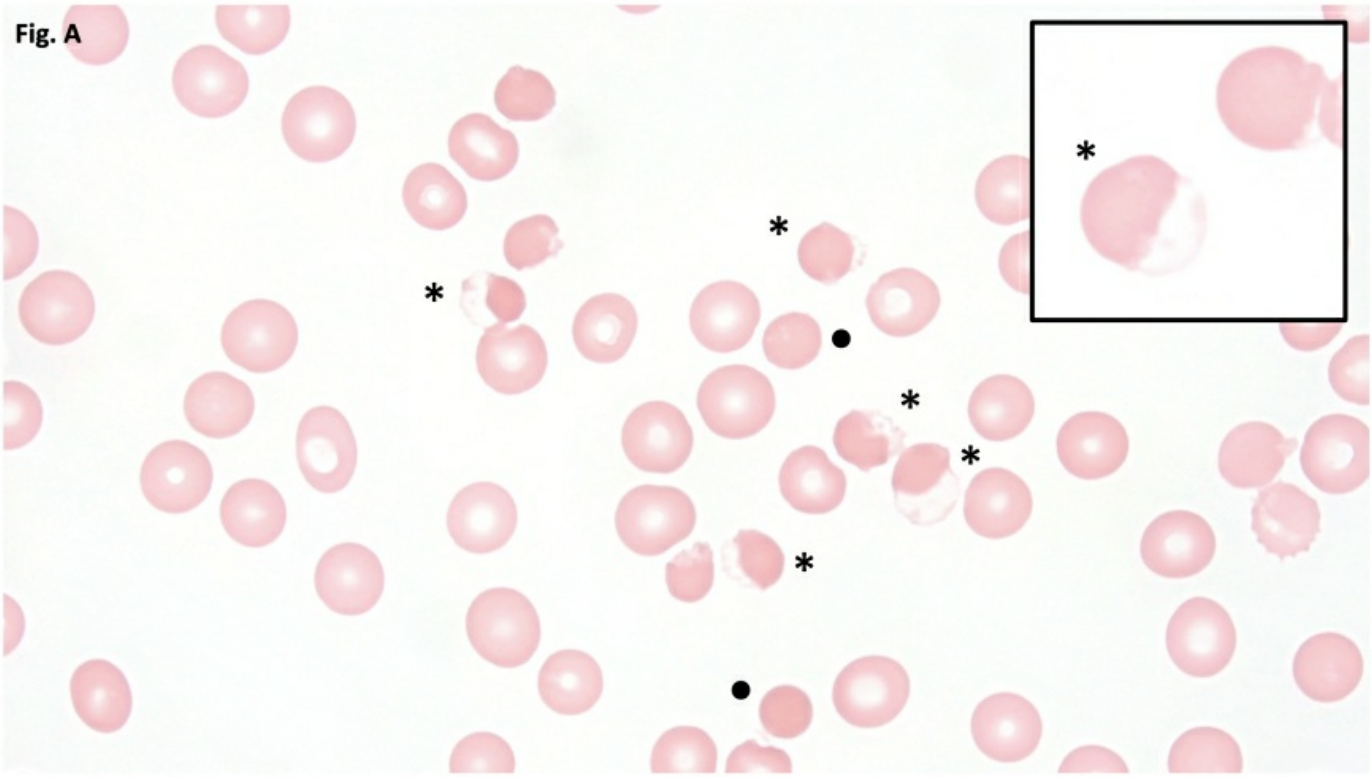

Fig. A. Oxidant hemolysis, peripheral blood findings. Multiple eccentrocytes (*), these cells show a peripheral defect covered by a thin residual membrane. Rare spherocytes (•) are also seen.

Her G6PD (glucose-6-phosphate dehydrogenase) screen came back deficient. She had episodes of severe dyspnea and slurred speech. However, her brain MRI was negative for acute stroke and her neurological status was attributed to anemic hypoxia. Her nadir hemoglobin level was 3.7 g/dl during her ICU stay. The patient declined any blood products, because of her religious conviction. Instead, the patient received supportive treatment with eltrombopag 150 mg/day, intravenous iron, erythropoietin 40,000 daily, ferric gluconate and folate until her counts recovered after transplant. Discussion of the novelty and importance of the specific case:Rasburicase is commonly used for treatment of hyperuricemia in patients with hematological malignancies. Rasburicase is an oxidant drug, and it well-known to cause severe hemolysis in patients with G6PD deficiency. This patient's rasburicase-induced hemolytic anemia occurred immediately after autologous stem cell transplant during severe myelosuppression. Management was further compromised by the inability to transfuse red cells. Conclusion:Rasburicase is associated with significant hemolysis in patients with G6PD deficiency and can lead to severe complications. All patients in the myeloma center are routinely screened for G6PD deficiency at intake, but this patient was missed. An electronic flag has been created in our medical record system to ensure that G6PD deficiency has been excluded before rasburicase is prescribed.

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