LETTER

An unusual case of hemolysis in a patient with cirrhosis

Dear Editor,

A 57-year-old man with decompensated alcoholic cirrhosis was admitted to the University Hospital of Heraklion with variceal bleeding. Hemostasis was achieved endoscopically by application of rubber bands and administration of sclerosing agents. He also received octreotide and four units of packed red blood cells (RBC) to maintain a hemoglobin level of 8 g/dl.

He had normal renal function on admission, and his liver function tests were abnormal with aspartate aminotransferase: 60 U/l, alanine aminotransferase: 38 U/l, total bilirubin: 10 mg/dl, and international normalized ratio: 1.5. The following days the clinical status of the patient improved. Upper gastrointestinal bleeding resolved and he remained stable with no further need for transfusions.

On the 15th hospital day he exhibited altered mental status, an abrupt fall in his hemoglobin level, a concomitant decline in his renal function with creatinine of 1.8 g/dl and a rising serum bilirubin level of 13.5 mg/dl (direct bilirubin: 8 mg/dl). Urgent endoscopy was negative for upper gastrointestinal (GI) bleeding.

He was transferred to the intensive care unit and during his stay there, his bilirubin level kept rising reaching 70 mg/dl with an indirect bilirubin level of 53 mg/dl. The patient became severely anemic and transfusion-dependent requiring 1-2 units of packed RBCs per day. He developed renal failure, and continuous hemodiafiltration was applied. Although the most obvious diagnosis was recurrent GI bleeding, the high levels of indirect bilirubin, which is a widely available and useful diagnostic index in these cases, helped us to include hemolytic anemia in our differential diagnosis. Laboratory studies revealed a non-immune mediated hemolytic anemia with negative direct antiglobulin test, a decreased haptoglobin level and elevated levels of lactate dehydrogenase. The peripheral blood smear showed a predominance of acanthocytes and target cells. The urine sediment was consistent with acute tubular necrosis probably as a result of pigment nephropathy due to hemolysis.

The working diagnosis was spur-cell hemolytic anemia (SCA). Over the following days, he developed multiple organ failure and died.

Severe hemolytic anemia is a rather rare phenomenon in patient with cirrhosis and is associated with the presence of spur cells[1]. Spur-cells are acanthocytes with morphological abnormalities due to the increase of cholesterol to protein and cholesterol to phospholipid ratios in the red cell membrane. Their spike-like projections deform their shape and flexibility, making them susceptible to trapping and destruction by the spleen[2,3]. Orthotopic liver transplantation has been shown to reverse spur cell anemia in adult patients with end-stage liver disease. Prognosis of patients with SCA is poor and the majority dies within months of diagnosis[1,3].

The clinical course of our patient correlates with the poor prognosis of this entity. Overall, SCA is a rare condition, predominantly seen in patients with advanced hepatocellular disease that should always be considered upon the development of hemolysis in this clinical setting.

References

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Conflict of interest

None.

Keywords: cirrhosis, hemolytic anemia, spur cells

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