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Letters to the Editor

Post-transfusion hyperhemolysis syndrome following gastrointestinal bleeding secondary to prehepatic portal hypertension

Key words: Hyperhemolysis syndrome. Gastrointestinal bleeding. Transfusion. Portal thrombosis.

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Dear Editor,

Hyperhemolysis syndrome (HHS) is a rare, life-threatening complication and early identification is key to improve prognosis (1).

Case report

A 30-year-old woman with portal cavernomatosis secondary to postnatal omphalitis and two bypass surgical procedures during childhood experienced multiple bleeding events from gastroesophageal varices. These required multiple transfusions and the antibody analysis was consistently irregular. At age 28, progressive thrombosis was identified in the inferior vena cava and other areas which responded to hypo-coagulation therapy. Eleven days before admission, the patient presented with self-limiting "sentinel" bleeding which required three red blood cells concentrates (RBCCs). The origin could not be identified due to the diagnostic limitations as the patient was pregnant. Following a curettage procedure due to a miscarriage, the patient was admitted due to a new bleeding event secondary to aberrant collateral circulation (duodenal pericholecystic varices on computed tomography and magnetic resonance imaging [CT/MRI], resulting in intermittent hemobilia). Five RBCCs were transfused with an almost null response. During admission, the patient had persistent severe anemia without re-bleeding, combined with fever, reticulocytopenia, hemolysis and a negative direct Coombs test. The patient started therapy with corticoste-

Table 1. Time course of laboratory parameters according to treatment

Day	*0**	*1#*	*2$*	*3¶*	*5Ω*	*7*	*9*	*14ϕ*	*32Ψ*
Hemoglobin (g/l)	60	74	72	63	57	72	77	88	103
Hematocrit (%)	19	21	22	19	18	23	26	29	38
Reticulocytes (x 10^9/l)		357				705			157
Total bilirubin (BR) (mg/dl)		4.1	1.9	3.5	1.3	0,6	0.5	0.4	0.4
Indirect BR (mg/dl)		3,3	1,1	2,5	0,6				
LDH (IU/l)		1,824	951	2,178		1,444	1,131	548	328
Haptoglobin (mg/dl)						0			18
Ferritin (µg/l)						604			137

*Pre-transfusion. #Post-transfusion, after five RBC concentrates. $Onset of IV methylprednisolone 1 mg/kg/12 h. ¶Onset of IV immunoglobulin 1 g/kg/24 hrs (five days). ΩOnset of IV iron sucrose (200 mg/48 h) and erythropoietin (EPO) 30,000 IU weekly. ϕHospital discharge on PO prednisone 60-0-30 mg, with descending dosage. ΨAt 30 days after steroid therapy onset.

roids and immunoglobulin. She was discharged after two weeks on a descending regimen of oral corticosteroids (Table 1) and required no further transfusions.

Discussion

HHS develops with no hematologic underlying disease in exceptional cases (2,3). It manifests with paradoxical post-transfusion decreased hematocrit (lysis of both transfused and host red blood cells), reticulocytopenia (rather than the expected reactive reticulocytosis), fever and evidence of intravascular hemolysis. Direct antiglobulin is negative in acute forms. Basic treatment includes steroids and immunoglobulin (3). Subsequent transfusions are both dangerous and ineffective, and therefore are initially contraindicated and should only be used in life threatening situations (4). Early clinical identification allowed this patient to receive specific therapy. Transfusions were discontinued and post-bleeding anemia (5) was managed according to laboratory parameters.

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