Management of a Patient with Bombay Blood Group and Chronic Liver Disease with Subdural Hematoma

Siddharth Chavali1, Subodh Raju1, Suresh Kanasani1, Abhirama C. Gabbita1

1 Institute of Neurosciences, AIG Hospitals, Gachibowli, Hyderabad, Telangana, India

Abstract

The Bombay blood group is an extremely rare entity within the conventional ABO blood grouping system. End-stage liver disease also presents with myriad disorders of coagulation due to impaired synthesis and dysfunction of clotting factors, which predisposes patients to spontaneous and life-threatening episodes of bleeding. We report a patient with Bombay blood group and end-stage liver disease who presented to our hospital with a spontaneous subdural hematoma. Although conventional parameters of coagulation in this patient were abnormal, we were able to safely defer product transfusion because his thromboelastography (TEG) report was within acceptable ranges. In this article, we discuss our strategy for optimization of extremely limited blood resources in this scenario and perioperative strategies for the management of coagulation anomalies in patients with liver dysfunction.

Keywords

► Bombay blood group
► Cirrhotic liver disease
► Subdural hematoma

Introduction

The Bombay blood group is an extremely rare autosomal recessive phenotype within the ABO blood group system, with an estimated prevalence of 1:10,000 in the Indian population and ~1:10,000,000 in the international population.1 Cirrhotic liver disease also presents with low platelet counts and coagulation factor deficiencies, placing them at an increased risk for spontaneous bleeding. We report a case with this Bombay blood group and chronic liver disease who presented to our hospital with a spontaneous subdural hematoma. The patient required emergency surgery, and perioperative concerns, as well as management, are discussed below.

Case Report

A 62-year-old male patient presented to our hospital with a history of holocranial headache for a week with increased lethargy for a day. His medical history was remarkable for a history of chronic liver disease (Child-Pugh Class C), hypertension, esophageal varices, and presence of the Bombay blood group. He was hemodynamically stable, and on neurological examination, he had a Glasgow coma score (GCS) of E3V4M6, bilaterally equally reacting pupils, and right-sided paucity of movement. On general examination, he was drowsy, icteric, exhibited flapping tremors, and had significant ascites. His initial laboratory studies revealed hemoglobin 12 g/dL, platelet count of 80,000, prothrombin time 23.0...
INR 1.89, and partial thromboplastin time (aPTT) of 48.5. Baseline thromboelastography (TEG) was done, which showed reaction (R) time of 5.2 minutes (2–8 minutes), maximum amplitude (MA) of 63 mm (51–69 mm), and lysis at 30 minutes (LY30) of 5% (0–8%). In the setting of normal serum ammonia levels, computed tomography (CT) imaging of the brain was performed that showed a large left-sided subdural hematoma (SDH) with mass effect, and a midline shift of ~7 mm. His blood group was reconfirmed with the previous treating institute as well as in our blood bank. A single unit of whole blood with this blood group was found to be available in the city, and this was kept on standby for any emergencies. The anticipated time required for the arrival of the whole blood unit was ~45 minutes, and this factor was borne in mind while planning the surgery.

Acute, clinically significant coagulopathy was ruled out in view of the TEG study findings, and he was planned for emergency evacuation of the SDH. On arrival in the operation theater, all standard monitoring was done as per the American Society of Anesthesiology (ASA) recommendations, and cannulation of the right radial artery was performed. He was sedated with dexmedetomidine at a rate of 0.5 µg/kg per hour targeting a Ramsay Sedation Scale score of 2–3, and scalp block was performed on the left side with 0.5% bupivacaine, targeting the supraorbital, supratrochlear, zygomaticotemporal, greater auricular, and greater and lesser occipital nerves. Tranexamic acid was given as a bolus of 15 mg/kg, followed by an infusion of 2 mg/kg per hour throughout the surgery. Burr hole evacuation of the SDH was performed, and the perioperative period remained uneventful. He also received perioperative supplementation of Vitamin K (10 mg IV) for 3 days. On postoperative day 3, an interval CT brain showed complete resolution of the SDH, and he was discharged in stable neurological condition on postoperative day 5.

**Discussion**

Acute SDH is a neurosurgical emergency often requiring urgent hematoma evacuation via a craniostomy. Liver cirrhosis itself may be a predisposing condition for falls, especially in patients with overt hepatic encephalopathy. Cirrhotic liver disease is associated with diffuse cerebral atrophy and increased incidence of arteriovenous malformations due to altered levels of pro-angiogenic factors. These changes lead to increased length as well as increased likelihood of bridging vein rupture, which may manifest as SDH formation.

Healthy individuals possess adequate amounts of clotting factors, fibrinogen, and platelets to achieve adequate clot initiation, propagation, and dissolution. However, patients with liver dysfunction have a disturbed balance of coagulation factors, causing deviation from the normal coagulation cascade. In these patients, decreased levels of all liver-dependent factors, such as vitamin K-dependent clotting factors (II, VII, IX, and X) may explain the prolonged INR. Additionally, functional platelet dysfunction, as well as thrombocytopenia secondary to splenic sequestration may also be a contributing aspect for decreased clot formation. These abnormalities can manifest as prolongations of PT and aPTT, which have led to this condition being considered a hemorrhagic disorder.

Bombay blood group (Oh or hh) was first described by Bhende et al in 1952. This blood group is an extremely rare autosomal recessive condition that occurs due to a point mutation of the gene encoding for H antigen on red blood cells. These patients lack A, B, and H antigens in their blood and produce anti-A, anti-B, and anti-H antibodies. Lack of A and B antigens leads to similarity with the O group, but the presence of anti-H antibody causes reactions with all blood types including the O group. Hence, these patients can only receive either autologous transfusion or blood from another person with the Bombay blood group as mismatched transfusion can lead to critical adverse events. Autologous blood transfusion can be a viable alternative for this patient population in the setting of elective surgeries, but in our case, this patient needed to be taken up on an emergency basis, and intraoperative blood loss was not predicted to be very high.

Thus, perioperative transfusion management with packed red blood cells is challenging given the concerns of a limited supply of available blood and the intraoperative need. Patients with this phenotype can receive fresh frozen plasma and cryoprecipitate as normal for the treatment of coagulopathies; however, platelet transfusion should be limited to type A2 because ABH antigens are expressed on platelets, and packed platelets may also contain a small number of donor RBCs. Given this conundrum, the use of viscoelastic tests of coagulation such as TEG may lead to a better assessment of functional coagulation status of these patients as compared with conventional assays, and lead to conservation of valuable blood products in this patient. Simply relying on conventional tests of coagulation in patients with liver disease may be an overly simplistic evaluation of a complex pathology. TEG seems to provide a more realistic assessment of in vivo hemostatic physiology, and its use can lead to more rational transfusion practices. Indeed, previous studies have also found that a TEG-guided transfusion strategy led to significantly lower use of blood components as compared with standard management (according to INR and platelet count) without differences in bleeding complications.

**Conclusion**

Patients with the Bombay blood group constitute a significant challenge because they are unable to receive blood components from any other phenotype. Liver cirrhosis in such patients further complicates the management in these patients due to altered coagulation parameters. Judicious use of components guided by point-of-care coagulation tests may help conserve vital blood components in such patients, for which a coordinated effort by the treating team and Bombay blood group organization is needed.

**Conflict of Interest**

None declared.
References
1 Oriol R, Candelier JJ, Mollicone R. Molecular genetics of H. Vox Sang 2000;78(Suppl 2):105–108