Emergency Laparoscopy For Intestinal Bleeding In A Patient With Glanzmann's Thrombasthenia

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Citation

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Abstract

Glanzmann's thrombasthenia (GT) is a rare disorder, which is characterized by abnormal platelet aggregation due to either absent or dysfunctional glycoprotein IIb-IIIa receptor. The patients are at risk of mucocutaneous bleeding, and excessive hemorrhage during surgery. This is a report of a patient with GT with abdominal bleeding, who presented for emergency laparoscopy, and resection of intestinal arteriovenous malformation. The patient was transfused both intraoperatively and postoperatively with packed red blood cells and platelets. The management of patients with GT presenting for surgery is discussed.

INTRODUCTION

Glanzmann's thrombasthenia (GT) is a rare disorder, which is characterized by abnormal platelet aggregation. It is an autosomal recessive disease, in which there is a functional impairment of platelets, due to either absent or dysfunctional glycoprotein IIb-IIIa receptor. These patients are at risk of mucocutaneous bleeding, and excessive hemorrhage during surgery. This is a report of a patient with GT with abdominal bleeding, who presented for emergency laparoscopy, and resection of intestinal arteriovenous malformation. To the best of my knowledge, this is the first report of a patient with GT, with internal bleeding, presenting for surgery.

CASE

The patient was a 70 year old man, with history of GT, who presented for emergency laparoscopy, and enterectomy for bleeding arteriovenous malformation (AVM) of the intestines. The patient had had surgery for intestinal resection for an AVM of the intestines 4 years earlier. That surgery had been complicated by bleeding, although more detailed information was not available. …He also had a history of ulcers in the duodenum, and stomach. Other medical conditions included hypertension, and hypercholesterolemia. His preoperative medical regimen included metoprolol, candesartan, avorvastatin, and esomeprazole. He had had preoperative bleeding, and was transfused 2u packed red blood cells (PRBC). The preoperative hematocrit was 23%. Due to the presence of antibodies to red cell antigens, the hospital did not have compatible blood for the patient, at the time of scheduling. The patient had antibodies to little E, JKA, and cell antigens. The surgery was delayed for 4 hours, until blood could be obtained from an outside blood bank.

In the operating room, 50 mg of diphenhydramine was administered intravenously. The patient then received 1 unit of washed and irradiated PRBC. Following uneventful anesthetic induction, and endotracheal intubation, general anesthesia was maintained with isoflurane, fentanyl, vecuronium, and oxygen. An arterial line was placed for continuous blood pressure measurement, and intermittent arterial blood sampling. The patient was transfused 1 unit washed, and irradiated PRBC intraoperatively, following a hematocrit measurement of 25%. The patient also received 1 unit of washed and irradiated single donor platelets during surgery, due to persistent oozing. A nasogastric tube was placed as per the request of the surgeon, which was atraumatic. The patient was extubated following the conclusion of the surgery. Postoperatively, the hematocrit was 24%, the PT INR 1.1, PTT 25, and platelet count was 137,000. The patient received an additional 2 U washed and irradiated PRBC postoperatively, and an additional single donor washed and irradiated platelets. On post operative day (POD) 3, the hemoglobin was 9.0gm/dl, the hematocrit was 27.2%, and the platelet count was 104,000. The patient was discharged home on POD5.

DISCUSSION

There is very limited information regarding patients with GT

presenting for surgery ([1,2]). This disease is a rare disorder, and is due to either absent or dysfunction glycoprotein (GP) IIb-IIIa. This results in a defect in platelet aggregation. Furthermore, these patients are often transfused both blood cells, and platelets, and may have developed antibodies to blood component antigens. Our patient had a history of multiple transfusions of both PRBC, and platelets. His type and cross revealed multiple antibodies, making it difficult to find compatible blood products. In fact, there were no compatible units in the hospital, and the surgery had to be delayed several hours until the blood products were obtained.

It may be very difficult to stop the bleeding in patients with GT ([1]). The platelets may not be effective at improving platelet function, if there are circulating anti GP IIb-IIIa antibodies. We used single donor platelets, which is preferable, and may be lifesaving ([3,4]). These were given intraoperatively, and postoperatively. Despite obtaining adequate surgical hemostasis, transfusion of both PRBC and single donor platelets were required postoperatively.

There was one prior report of a patient with GT undergoing abdominal surgery ([1]). A 21 year old man with GT, with abdominal pain and distention, presented for emergency laparotomy. As in our case, platelets were administered prior to incision. These authors reported the use of 3 ABO identical plateletpheresis units. In addition, rF VIIa 90 ug/kg (Novoseven, Novo Nordisk, Denmark) was administered. Although the bleeding time decreased from over 30 minutes to 18 minutes following the transfusion of platelets, the platelet count did not increase. An additional 3 units platelets were administered during surgery. Due to bleeding at the NG tube insertion site, the patient was kept intubated at the conclusion of surgery, and was transferred to the intensive care unit (ICU). Although the nose was packed, there was persistent nasopharyngeal bleeding, and the patient received multiple doses of rFVIIa postoperatively through post op day (POD) 1. The patient also received 6 units of platelets postoperatively.

There has been one report of a patient with GT undergoing cardiac surgery ([2]). This patient underwent coronary artery bypass grafting (CABG). This patient received single donor platelets preoperatively, intraoperatively upon separating from cardiopulmonary bypass, and postoperatively. The patient required exploratory sternotomy on POD2 for release of cardiac tamponade. There was no active bleeding site identified at that time. That patient had received a total of 25 units PRBC, 12 units fresh frozen plasma, 60 units of random donor platelets, and 25 units single donor platelets. The platelet count gradually recovered to normal over 5 days.

In summary, we report for the first time, a patient with GT presenting for surgery for treatment of internal bleeding. Since there may have been alloimmunization to blood products, it is important to consult preoperatively with the blood bank, to ensure the availability of blood and platelets. We recommend the use of single donor platelets, for treatment of platelet dysfunction. The use of rFVIIa should be considered, if platelets are ineffective, or can even be considered as first line treatment when there are circulation antibodies to platelets.

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