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ANAESTHETIC MANAGEMENT FOR EMERGENCY LAPAROTOMY IN A PATIENT OF GLANZMANN THROMBASTHENIA

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ABSTRACT Glanzmann thrombasthenia (GT) is a rare disease of an autosomal recessive inheritance characterized by a fatal bleeding tendency. The anaesthesiologist should be cognizant of the risk involved and be prepared with necessary measures. This paper presents the successful management of intussusception in a 7-year-old with GT undergoing emergency laparotomy in a resource-limited setup where thromboelastography and recombinant factor VIIa (rFVIIa) are not available.

KEYWORDS : Glanzmann thrombasthenia, anaesthesia, emergency, laparotomy

INTRODUCTION

Glanzmann thrombasthenia (GT) is a rare (1/1,000,000) disorder with autosomal recessive inheritance and is characterized by deficiency or functional defect of glycoprotein IIb/IIIa (GP IIb/IIIa), which acts as a fibrinogen receptor on the thrombocyte surface. GP IIb/IIIa receptors play a basic role in thrombocyte adherence and aggregation. Thrombocytes of such patients fail to bind fibrinogen and aggregation does not occur. Patients are at a high lifetime risk of severe bleeding, particularly during surgical procedures. Typically, such patients have a normal thrombocyte count, normal prothrombin time, normal partial prothrombin time, prolonged bleeding time, and impaired platelet aggregation (prolonged modified clot retraction).

The disease is usually diagnosed in young age after epistaxis or mucocutaneous bleeding. Complaints such as easy bruising, muscle hematomas, haemarthrosis, gastrointestinal bleeding, menorrhagia, and haematuria appear in further stages of life. The disease has no specific treatment. Such patients require a specific treatment regimen in the perioperative period for adequate functioning of the coagulation system.

Case Report

A 7-year-old boy patient (30 kg) was presented in the emergency department with complain of acute onset of abdominal pain and vomiting. On investigation, a large ileo-colic intussusception with mild ascites was diagnosed for which emergency laparotomy was indicated. On eliciting history from parents and reviewing the previous medical records, it was revealed that the patient was a diagnosed case of GT since the age of 2 years. Patient had multiple episodes of uncontrolled nose bleeding and gum bleeding in the past. He was treated with tranexamic acid during those acute bleeding episodes. Family history of the patient revealed that the mother and sister also suffer from the same condition. Preoperative haematological laboratory findings revealed a thrombocyte level of 461,000 μL^{-1}, haemoglobin level of 6.3 g dL^{-1}, activated partial thromboplastin time (aPTT) of 24.2 seconds, international normalized ratio (INR) of 1.08, and white blood cell (WBC) count of 12,740 μL^{-1}. Hepatic and renal function tests were within the normal ranges.

After standard monitorization (pulse oximeter, electrocardiography, non-invasive arterial blood pressure, body temperature) of the patient, who was transferred to the operating room, peripheral vascular access was established on the dorsal aspect of the right hand using 22-gauge cannula. Midazolam (1 mg), fentanyl (30 µg), propofol (60 mg) and succinylcholine (30 mg) were administered for anaesthesia induction. No complication was encountered during intubation procedure. Considering probability of bleeding after intubation, additional peripheral vascular access was established on the dorsal aspect of the left hand using a 22-gauge cannula. Maintenance of anaesthesia was provided by dexmedetomidine infusion (1-2 µg kg^{-1} $hour^{-1}$) and 0.5–1 MAC (minimum alveolar concentration) sevoflurane. Intraoperative blood loss was 400 mL, without any episode of incessant bleeding. Intraoperatively, two packed red blood cells (leukoreduced to prevent alloimmunization) were transfused. Tranexamic acid 1 g intravenous (IV) was administered before the commencement of surgery followed by 1 g infusion over 8 hours. The surgery lasted for 120 minutes. At the end of the procedure, residual neuromuscular blockade was antagonized with neostigmine 50 µg/kg and glycopyrrolate 10 µg/kg, and the patient's trachea was extubated with minimal evidence of gum bleeding. The patient was shifted to pediatric ICU for monitoring.

In the first 24 hours after surgery, the patient remained hemodynamically stable and the wound remained dry and clean. He was discharged on post-operative 8th day without any complications.

CONCLUSION

GT is a rare platelet disorder with an incidence of 1 in 10,00,000, with sparse literature available on the perioperative management, especially in emergency situations. Patients with GT are at risk of excessive bleeding during surgery or after trauma. Available therapeutic options are rFVIIa therapy and antifibrinolytics. Although rFVIIa is recommended as the first-line therapy for patients with GT undergoing surgery, its high cost compels us to consider other treatment modalities. Administration of tranexamic acid was found to be a cheap and effective alternative to control bleeding in our patient. Herein, we intend to highlight that perioperative platelet transfusion and tranexamic acid prophylactically may reduce intraoperative and postoperative bleeding in places where rFVIIa and monitoring facilities like TEG are unavailable.

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