Case Report

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Simultaneous occurrence of intussusception and duodenal haematoma in a patient with Glanzmann's thrombasthenia

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ABSTRACT

Glanzmann's thrombasthenia (GT) is a rare autosomal recessive disorder in which the platelets have anomalies of fibrinogen receptors causing bleeding tendencies. The disease usually presents with mucosal bleeding, petechial haemorrhages or gastrointestinal (GI) bleeding. Only sporadic cases of intussusception or duodenal haematoma have been reported with GT. We present a case of 5-year-old girl admitted with non-bilious vomiting, watery diarrhoea and abdominal pain. She is a known patient of Glanzmann's thrombasthenia.

Key words: Complications, duodenal haematoma, Glanzmann thrombasthenia, intussusception

INTRODUCTION

Glanzmann's thrombasthenia is an extremely rare autosomal recessive disorder caused by qualitative or quantitative deficiency of fibrinogen receptors (αIIbβ3) on the surface of the platelets. The incidence of the disease is about 1:1,000,000.[1] The gene for this receptor is located on chromosome 17 and any defect in the gene can lead to failure of platelets aggregation and hence prolonged bleeding time.[2] Glanzmann thrombasthenia (GT) has been classified into Type-I to Type-III on the basis of severity, deficiency of receptors, their functions and expressions. Our patient falls under Type-II as per our laboratory investigations.[3] The essentials for diagnosis include normal platelet count and morphology, prolonged bleeding time but normal prothrombin time and activated partial thromboplastin time with the absent platelet aggregation.[4] Diagnosis is usually by flow cytometry and DNA analysis.

Intussusception is a known complication of coagulopathies such as idiopathic thrombocytopenic purpura (ITP).[5] The occurrence of intussusception in a patient with Glanzmann's thrombasthenia has been reported rarely. Moreover, the association of intussusception and duodenal haematoma in the same patient is very rare and not reported in the available literature. This case report describes such an association and the effective surgical as well as medical management at our hospital.

CASE REPORT

A 5-year girl presented to emergency department with complaints of colicky abdominal pain, non-bilious vomiting and watery diarrhoea for the past 2 days. She was diagnosed as having gastroenteritis and was treated for moderate dehydration as an inpatient. She is a known patient of Glanzmann's thrombasthenia with a history of recurrent epistaxis and easy skin bruising which was followed up in other hospital.

Initial blood investigations including complete blood count, platelet counts, prothrombin time, activated

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partial thromboplastin time and electrolytes were within normal limits. An ultrasound performed on the next day suggested intussusception with incidental findings of the absent left kidney. After resuscitation and transfusion of blood products and recombinant activated factor VII (rFVIIa), laparoscopy was performed. It showed about 20 cm long ileoileal intussusception [Figure 1]. Patchy haemorrhagic areas were noted on the intestine. Reduction was achieved without much difficulty. Easy bruising of the intestines was noted during the laparoscopic procedure. The child was infused clotting factors and fresh frozen plasma as per advice of the treating paediatricians. The child improved and a repeat ultrasound done on the next day showed thickening of the small bowel loops without any recurrence of intussusception.

On the 2nd post-operative day, she started vomiting streaks of blood and later passed melaena. X-ray abdomen showed minimal central abdominal gas and also no gas in the rectum suggestive of upper gastrointestinal (GI) obstruction. Blood investigations showed very high levels of serum amylase and serum lipase. The child was treated conservatively and kept nil per oral. A repeat ultrasound on the 4th post-operative day showed a mass in the second part of the duodenum suggestive of duodenal haematoma. Magnetic resonance imaging (MRI) was performed which was also suggestive of duodenal haematoma with complete occlusion of the lumen [Figure 2]. TC99M isotope tagged red blood cell (RBC) scan performed on the same day showed active bleeding in the epigastric region. Selective angiography of gastroduodenal vessels was then performed with a plan to embolise the bleeding vessels, but selective angiography not shows any active bleeding [Figure 3] and embolisation was, therefore, deferred. At the same time, peripherally inserted central catheter line insertion was done for rFVIIa infusions and total parenteral nutrition. The child was kept on parenteral nutrition and was vomiting after taking even a small amount of oral fluids. On the 7th post-operative day, she vomited about 120 ml of clotted blood, which was probably spontaneous drainage of the duodenal haematoma. She immediately showed signs of improvement and started tolerating oral fluids. Her amylase and lipase level gradually returned to normal levels. Increasing dose of rFVIIa and a unit of packed RBC were given.

On day 12, she developed fever with epistaxis. She was evaluated for the focus of infection and found to have right lower zone haziness in the lungs. Her blood and urine culture were negative, but there was rise in inflammatory markers. Later, the patient was shifted to other hospital in view of sepsis and specialised Intensive Care Unit (ICU) care. She was found to have endocarditis being the reason for sepsis. She was intubated and was in the ICU for almost 3 weeks and later had a full recovery. She is now awaiting further treatment for gestational diabetes (GD).

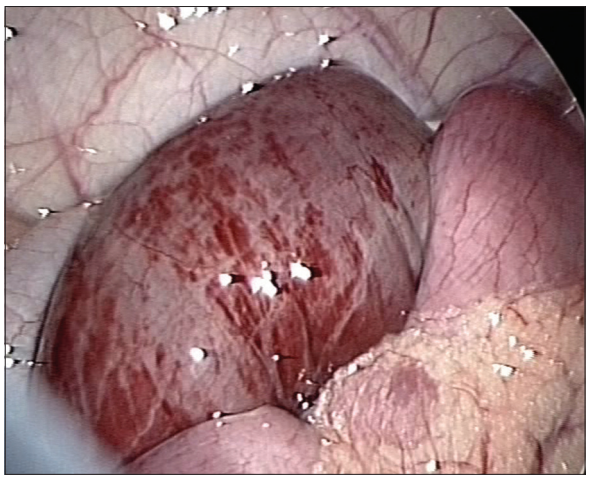

Figure 1: Ileoileal intussusception with haemorrhage on laparoscopy

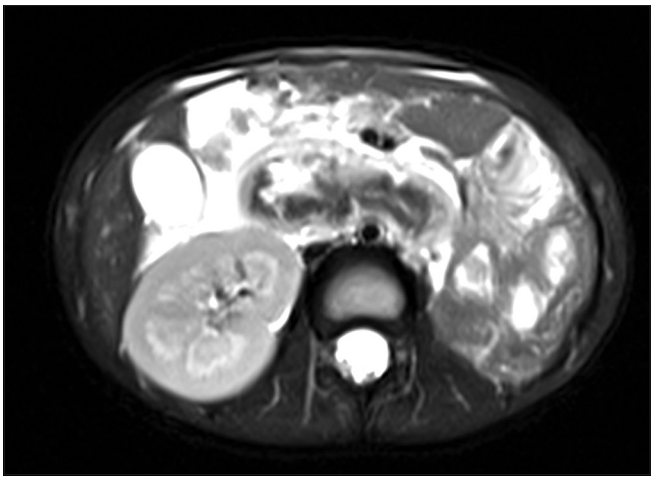

Figure 2: Magnetic resonance imaging showing duodenal haematoma

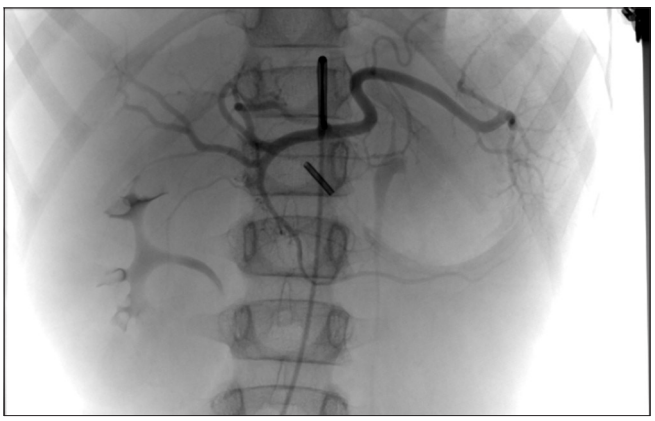

Figure 3: Angiography showing minimal extravasation

DISCUSSION

Dr. Eduardo Glanzmann first described the disease in 1918.[2] GD has an autosomal recessive inheritance. The disease is characterised by mucosal bleeding, easy bruising, excessive bleeding during circumcision, GI bleeding and menorrhagia.[3] The GT has been classified into Type-I to Type-III on the basis of severity, deficiency of receptors, their functions and expressions, however, there is no definitive correlation between the type of GD and the severity of bleeding, i.e., patients with nearly absent glycoprotein receptors can have minimal bleeding compared to patients who have good number of receptors.[4] The patients may remain symptom-free for many years in between episodes of bleeding. The GI complications are usually preceded by mucosal bleedings. Our patient also had history of epistaxis twice before this admission. GI bleeding usually results in upper or lower GI bleeding but can also result in duodenal haematoma, which causes abdominal pain and signs of upper intestinal obstruction.[5] Duodenal haematoma can occur spontaneously without any history of abdominal trauma in patients with GT, or they may be associated with the history of trauma.[5,6] Only a few cases have been reported for both the entities. Similarly, intussusception has been frequently reported in other coagulopathies, such as ITP and Henoch–Schonlein Purpura, however, its association with GT is rare and only a few cases have been reported.[7,8] Combination of intussusception and duodenal haematoma in the same patient has not been reported in the available literature. Our patient presented with intussusception as well as duodenal haematoma, which is a rare combination of complications associated with GT. The patient initially presented with intussusception and we thought there may be a possibility of iatrogenic injury during the laparoscopic reduction of intussusception causing duodenal haematoma; however, the procedure was smooth and extra cautions were taken to handle the intestine as the child was known case of GT. Laparoscopy gives better results for reduction of intussusception due to less tissue trauma.[8] Also looking back at the video recordings of the procedure, we could not find any indication of an iatrogenic injury.

Studies have shown that most of this haematoma can be managed conservatively and very rarely need intervention. Touloukian has clearly mentioned in their studies that non-operative management with rFVIIa treatment is highly beneficial in duodenal haematoma patients.[9] In July 2014, the Food and Drug Administration approved rFVIIa as the first recombinant treatment for bleeding episodes and perioperative management in patients with Glanzmann's thrombasthenia refractory to platelet transfusions, with or without antibodies to platelets.[10] Our patient also received rFVIIa post-operatively in increasing doses, however, the symptoms persisted and as the symptoms continued, we performed angiography to exclude and treat an active bleed in the duodenum. No active bleeding was noticed supporting the recommendation that conservative treatment shall be effective in most patients. It is also interesting to note that once the duodenal haematomas were evacuated spontaneously, the child improved. This highlights the fact that until the haematoma is drained or resolved the child will continue to be symptomatic. Therefore, the child with duodenal haematoma even though symptomatic may not have active bleeding and will need some time for the haematoma to resolve or drain spontaneously. During this period, the child will need parenteral nutrition. The parenteral nutrition also has its own side effects, and outpatient developed central line sepsis and needed aggressive management to treat these complications.

CONCLUSION

This case report defines the rare occurrence of both intussusception and duodenal haematoma in a patient with Glanzmann's thrombasthenia. Duodenal haematoma shall be considered in a coagulation disorder patient who is presenting with abdominal pain and vomiting. Ultrasound is the prime modality of investigation although MRI can be an adjuvant in the diagnosis. Most patients with duodenal haematoma can be managed conservatively with blood products and rFVIIa, however, few may need surgical intervention.

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Conflicts of interest
There are no conflicts of interest.

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