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Cardiac surgery in a patient with Glanzmann's thrombasthenia

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ABSTRACT

Cardiac surgery in patients with bleeding disorders like Glanzmann's thrombasthenia is quite challenging because of the increased risk of postoperative bleeding resulting in higher transfusions and associated morbidity. Platelet transfusion and use of antifibrinolytic drugs help in achieving optimum haemostasis in the patients with Glanzmann's thrombasthenia. Here, a case of six-year-old boy with Glanzmann's thrombasthenia is reported, who underwent surgical closure of atrial septal defect and was managed with platelet transfusion and tranexamic acid. The patient recovered well without having significant bleeding in the post-operative period.

Keywords: Atrial septal defect; cardiac surgery; congenital heart disease; Glanzmann's thrombasthenia; platelet disorder.

INTRODUCTION

Glanzmann's thrombasthenia (GT) is characterised by abnormal platelet aggregation, clot retraction and increased bleeding tendency. The risk of bleeding increases significantly during trauma or surgery. Non-cardiac surgeries have been performed in these patients with management of bleeding by using platelet, aminocaproic acid, tranexamic acid and recombinant factor seven transfusion.[1] A few case reports of cardiac surgery being performed in these group of patients are also available from other countries.[2] All of these cases were managed by a multidisciplinary team of cardiac surgeons, haematologists, and intensivists to ensure a reduction in postoperative blood loss allowing a smooth post-operative recovery.

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CASE REPORT

A six-year-old boy with history of intermittent epistaxis since the age of nine months, was incidentally diagnosed to have a large ostium secundum atrial septal defect (ASD) and referred to our center for surgical closure of the defect. The patient was investigated for bleeding disorder in the past. He was found to have homozygous ITGB3 gene mutation in exon five and confirmed to have Glanzmann's thrombasthenia. Since then, he was under regular care of pediatricians and haematologists for repeated epistaxis and haemoptysis. He had recurrent hospital admissions in the past for low haemoglobin and low blood pressure where he was managed with blood and platelets transfusion and oral iron supplemention. It was in one of the recent hospital admissions, when he was incidentally diagnosed to have an atrial septal defect.

On arrival to the study site hospital, his blood pressure was 96/50 mmHg, heart rate of 106 per minute, respiratory rate of 24 breaths per minute and

temperature of 98.2°F. His general physical examination did not reveal any petechiae, ecchymosis or haematoma. The cardiovascular system examination revealed a fixed wide splitting of the second heart sound. His respiratory system examination was normal.

Chest X-ray demonstrated cardiomegaly with hyperemic lung fields. Echocardiogram showed a 2cm x 2cm ostium secundum ASD with absent postero-inferior and inferior vena caval rims, dilated right atrium and ventricle, normal biventricular function (Figure 1). His preoperative blood investigation revealed haemoglobin level of 13.1gm/dl, platelet count of 188,500/mm^3, normal liver and renal function tests. Haematologist advised to manage the intraoperative and postoperative bleeding with blood and platelet transfusion, administration of tranexamic acid and Factor VII.

The patient underwent surgical ASD closure via median sternotomy followed by aorto-bicaval cannulation, initiation of cardiopulmonary bypass (CPB) and cardioplegic arrest of the heart in diastole. On performing a right atriotomy, an ostium secundum ASD with a diameter of approximately 2cm was visualised (Figure 2). The ASD was closed using autologous pericardial patch (Figure 3) and CPB was weaned off uneventfully. The aortic cross clamp time was 17 minutes and CPB time was 33 minutes. He had received 500 milligrams of tranexamic acid injection at the time of induction. Despite, minimal intraoperative blood loss, the haemoglobin was 9.1 gm/dl at the time of termination of cardiopulmonary bypass. So, he was transfused with 300ml of packed red cells. Patient was extubated after two and half hours of transferring to intensive care unit. The total volume of postoperative drain was one hundred and seventy milliliters on the first twenty-four hours of surgery followed by twenty-five milliliters in next two days. He had multiple episodes of haematemesis on second and third post-operative days, with a drop in haemoglobin to 6 gm/dl. He was managed in intensive care unit with three transfusions of 15ml/kg of packed cells, one 15ml/kg of platelet and injectable tranexamic acid 250 mg three times a day for five days. Factor VII was not used, as it is not available in the country. He did well later in the post-operative period and was discharged from hospital on sixth post-operative day.

He was readmitted to hospital after seven days of discharge for fever and pneumonia. He was treated with intravenous antibiotics and discharged from hospital in one week. Echocardiogram at discharge revealed an intact ASD patch without residual leak and normal biventricular function.

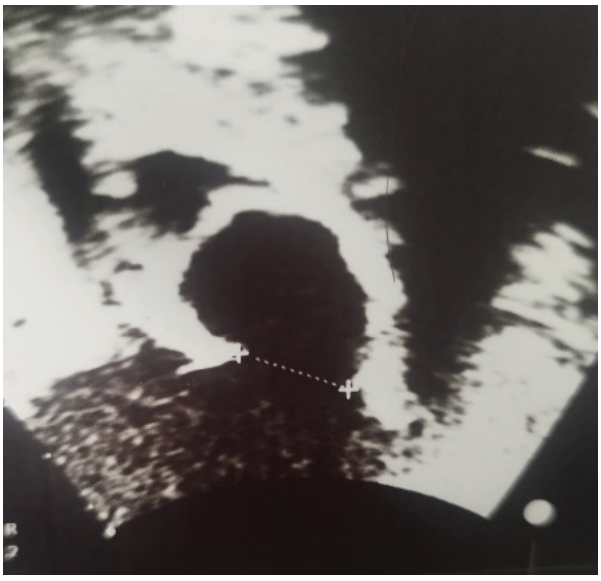

Figure 1: Echocardiogram picture showing an ostium secundum atrial septal defect

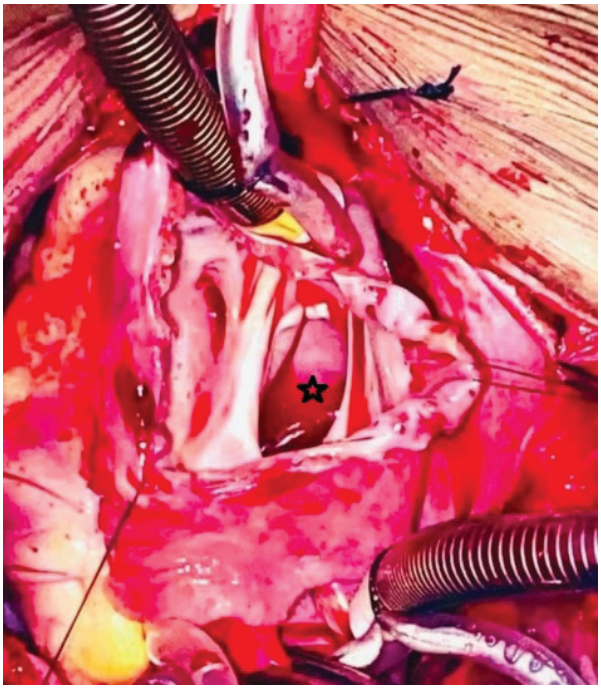

Figure 2: Intraoperative picture showing atrial septal defect (marked with a star)

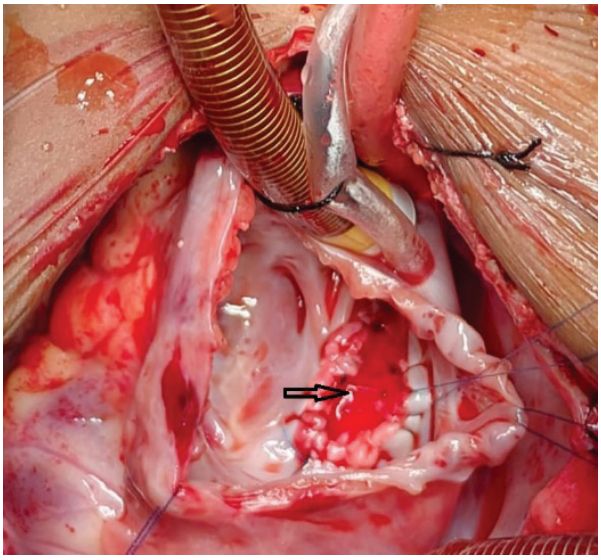

Figure 3: Intraoperative picture showing ASD closure with pericardial patch (marked with an arrow)

DISCUSSION

Eduard Glanzmann first described this hereditary haemorrhagic thrombasthenia in 1918 A.D.[2] Glanzmann's thrombasthenia is an autosomal recessive bleeding disorder which is characterised by mutation in ITGA2B and ITGB3 genes in chromosome 17 leading to abnormality of glycoprotein IIa / IIIb receptor and impaired platelet aggregation and haemostasis.[3] The acquired form of this disease is caused by antibodies to platelet αIIbβ3.[4] Clinical manifestation of GT include petechiae, purpura, gum bleeding, epistaxis, bleeding from gastrointestinal tract, menorrhagia and rarely haemarthroses. These symptoms along with the laboratory parameters like normal platelet count, prothrombin time and activated partial thromboplastin time, increased bleeding time and decreased platelet aggregation in response to various factors like collagen, epinephrine, adenosine diphosphate and normal platelet aggregation in response to ristocetin and genetic testing confirm the diagnosis.[1] The bleeding episodes in GT are managed with platelet transfusion, recombinant Factor VII, tranexamic acid. However, repeated transfusion of platelets can lead to alloimmunisation and non-responsiveness to platelet transfusion. Therefore, any surgery in GT is associated with increased incidence of bleeding.

The risk is higher in cardiac surgery with the use of cardiopulmonary bypass. Higher doses of heparin, cardiopulmonary bypass related shear stress, platelet dysfunction, activation of fibrinolysis accentuate the bleeding tendency in such patients. There are not many reported cases of cardiac surgery in GT patients. Simha et al., described performing mitral valve repair in a patient with GT and the perioperative bleeding was managed with transfusion of packed cells, single donor platelet and avoidance of non-steroidal anti-inflammatory drugs and antiplatelets.[2] Seikh et al., reported open aortic valve replacement, with transfusion of thirty-five apheresed platelets and administration of 1-deamino-8-d-arginine vasopressin (DDAVP), infusion of aminocaproic acid, for the management of postoperative bleeding.[3] Another paper describes successful treatment of postoperative bleeding following coronary artery bypass grafting with tranexamic acid infusion and autologous blood transfusion.[5] Other reports describe ventricular septal defect closure with tricuspid valve replacement and redo mitral valve replacement in patients with GT.[6,7] Truong et al., highlighted the safety of short term dual antiplatelet drugs for one month followed by maintenance clopidogrel monotherapy, in a patient following coronary artery stenting, as dual antiplatelet therapy in patients with GT can cause fatal bleeding.[8]

In our setting, the blood products like apheresed platelets, irradiated platelets are not available. Similarly, the recombinant factor VII, aminocaproic acid and injectable desmopressin are also not available in Nepal as of now. Therefore, managing perioperative bleeding following a cardiac surgical procedure in GT patients, in low resource setup like ours, depends solely on the availability of platelet rich plasma, platelet concentrate and tranexamic acid in addition to the packed red blood cells. Our patient also did well with platelet and packed cell transfusion and administration of tranexamic acid.

CONCLUSION

Glanzmann's thrombasthenia and increased bleeding pose extra challenge to cardiac surgical procedures. There are no specific guidelines on the management of these patients. In low resource countries, adequate blood and blood products transfusion and use of easily available antifibrinolytic agent that is tranexamic acid, help in reducing incidence of significant post-operative surgical bleeding.

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