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ANAESTHETIC CONSIDERATIONS IN AN OBSTETRIC PATIENT WITH GLANZMANN'S THROMBASTHENIA POSTED FOR EMERGENCY CAESAREAN SECTION: A CASE REPORT

Anaesthesiology

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ABSTRACT

Glanzmann's Thrombasthenia, a rare inherited autosomal recessive bleeding disorder of platelet function characterized by multiple bleeding episodes. It is hazardous for these patients to conceive and undergo surgical intervention with a high risk for antepartum and postpartum haemorrhage. We describe anaesthetic considerations of a 29-year primipara patient with Glanzmann's Thrombasthenia presented with 9 months of amenorrhea for emergency caesarean section. Caesarean section was successfully managed with help of TEG with no bleeding complication during and after the procedure and the patient was successfully discharged on the fourth postoperative day.

KEYWORDS

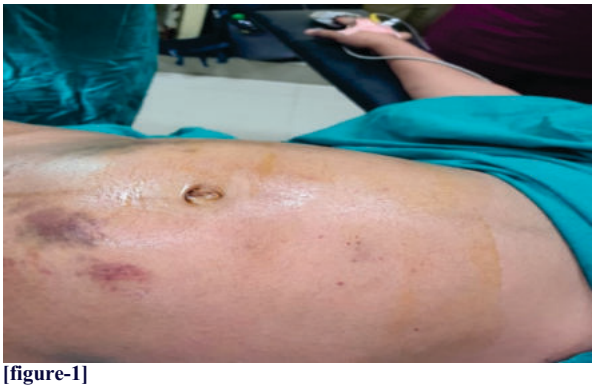
Glanzmann's Thrombasthenia, Obstetric patient, Thromboelastography

INTRODUCTION

Glanzmann's thrombasthenia (GT) is a rare inherited autosomal recessive platelet disorder manifested by abnormal GP 2B/3A Complex on platelet membrane, which acts as a receptor for fibrinogen during the event of platelet aggregation. This disorder is characterized by abnormal platelet function, resulting in prolonged bleeding time, prolonged closure time on platelet function analyser (PFA-100) [1] despite normal platelet count, normal PT/INR, and APTT. GT manifests as mucocutaneous bleeding, menorrhagia, bruising to trivial trauma since early childhood.[2] Hemarthrosis, muscle hematoma, haematuria, and serious GIT bleeding are rare. These patients are at high risk for life-threatening bleeding following major surgery in perioperative bleeding. Major bleeding requires blood and blood component transfusion and antifibrinolytic agents. The special measure includes Recombinant Factor 7A, allogenic stem cell transplant, and gene therapy.[3]

CASE REPORT

A 29-year full-term primipara presented to the obstetric department with a complaint of abdominal pain. On obstetric evaluation, it was found to be fetal bradycardia and the patient was scheduled for an emergency caesarean section. History revealed that she is a known case of Glanzmann's Thrombasthenia since childhood. She had menorrhagia lasting for 15-20 days during menarche and had occasional episodes of epistaxis with history of 4-5 episodes of blood and blood product transfusion to date. Family history revealed brother diagnosed with GT having history of frequent epistaxis and gum bleeding, no other family member diagnosed with GT or having consanguineous marriage. Obstetric history revealed, missed abortion at 2 months of gestation, 3 years back not associated with any increased bleeding episode. On general examination pallor was present, and petechiae were present in both lower limb. On abdominal inspection, there were multiple purpuric patches in lower abdomen. [figure-1]



[figure-1]

Haematological investigation showed haemoglobin of 9g/dl, platelet count of 1.55 lacks/cumm, PT of 9.2 seconds, INR 0.79, aPTT of 27.6, Bleeding time was6 minutes clotting time was 13 minutes. Blood group was B positive. All other investigations were normal. Blood sample was sent for thromboelastography. Six units of whole blood was cross-matched, request for Single donor platelet was sent and six units of random donor plasma was arranged. On shifting the patient to OT, venous access was secured by two 16G iv cannula on the cubital fossa. ASA standard Monitors were attached, and baseline vitals were recorded. RDP transfusion started. General anaesthesia was planned, inj.metoclopramide 10 mg iv, inj. ranitidine 50 mg iv given. A rapid sequence induction of anaesthesia was performed with inj. propofol 150 mg iv and inj. succinylcholine 100 mg iv, followed by gentle endotracheal intubation with a 7.0 mm cuffed endotracheal tube, avoiding any trauma to the mucosa. Anaesthesia was maintained with O2, N2O, sevoflurane, and inj. atracurium 20 mg iv loading followed by 5 mg iv intermittently. A bolus of 1 IU iv followed by an infusion of 4-5 IU/hour of inj. oxytocin was administered after delivery of the baby, inj. fentanyl 100 µg iv given iv and inj. paracetamol 1g iv was given for analgesia. Inj. tranexamic 1 g I.V. given over 10 min. [1,5] TEG report showed reduced alpha angle and maximum amplitude. Second sample for TEG was sent. Caesarean section progressed uneventfully, with a blood loss of 800–1000 ml. There was cardiovascular stability throughout the procedure. Meticulous haemostasis was achieved before wound closure. Residual neuromuscular blockade was reversed with inj. glycopyrrolate 0.4 mg iv and inj. neostigmine 2.5 mg iv and the patient's trachea was extubated. A single intraperitoneal drain was inserted by surgeon intraoperatively.

Second TEG report was normal. Postoperative investigation showed haemoglobin of 8 g/dl, platelets 1.34 L/cumm. One-unit PCV was infused and Inj. Tranexamic acid 10 mg/kg 8 hourly was continued in postoperative period. Abdominal drain output was 100 ml in first 12 hrs. and 200 ml in next 24 hrs and was removed. The postoperative period was uneventful with patient maintaining vitals within normal limits, adequate urine output, no postpartum haemorrhage. The patient was discharged successfully after 3 days and called up for follow-up after 7 days.

DISCUSSION

Eduard Glanzmann, a Swiss pediatrician, first described this disease in 1918 as “hereditary hemorrhagic thrombasthenia”.[4] GT is the most frequent inherited disorder of platelet receptor integrin complex glycoprotein (GP) IIb and IIIa. GT classified into Quantitative (type 1 and 2) and Qualitative disease (type 3). Type I disease, it is most severe form of GT with GP 2B/3A Levels < 5%. Type II has 5–15% expressed levels of GP 2B/3A and Type III are heterozygous carriers with normal levels of dysfunctional GP IIb/IIIa levels.[4,7,10] Diagnosis of GT is

based on history, mucocutaneous bleeds, purpura, uncontrolled bleeding after major surgery, bleeding from trivial trauma, laboratory tests include- absence of aggregation response of platelets to all platelet agonists such as ADP, collagen, arachidonic acid, and ristocetin. Platelet adhesion in GT is normal. TEG analysis will show normal clot initiation and poor clot strength in patients with GT which can guide regarding platelet transfusion.[10] In GT patients due to history of multiple transfusion there is high chance of antiplatelet antibody production this may cause platelet transfusion ineffective keeping this in mindand to avoid probable complications associated with neuraxial blocks like epidural hematoma and to have better control of intraoperative hemodynamic we chose general anaesthesia.[3,7] Platelet transfusion is standard management for GT [4] as it was an emergency case 6 units of RDP [6] were transfused, making arrangements for SDP. Additional coverage was given with antifibrinolytic drug, Tranexamic acid, acting through inhibition of plasminogen conversion to plasmin and subsequent fibrinolysis inhibition. [2,8] topical fibrin glue was instilled intraperitoneally after uterine closure for additional hemostasis. [9] Proper surgical hemostasis and oxytocic's also play important role for control of bleeding. [1,6] Postoperative period was managed with infusion of one unit of PCV and one unit of SDP. SDP were used to counteract alloimmunization by repeated platelet transfusion and resultant alloantibody mediated donor platelet destruction. [2,7]

CONCLUSION

Successful outcome of GT patients undergoing major surgical procedures can be achieved by a multidisciplinary team approach involving surgeon, anesthesiologist and hematologist. TEG plays a crucial role decision making regarding platelet transfusion and patient management.

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