University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2019 , Vol. 5(6)

Anaesthetic management of term pregnant patient Glanzmann thrombaesthenia

THIRUVARUL SANTHOSHINI
Department of Anaesthesiology,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : Glanzmann thrombaesthenia is a rare autosomal recessive bleeding disorder caused by deficient or dysfunctional glycoprotein IIb-IIIa receptor on platelet membrane resulting in defective platelet aggregation. We report a case of Glanzmann thrombaesthenia undergoing lower segment Caesarean section and discuss about its perioperative management.

Keyword : Glanzmann thrombaesthenia, general anaesthenia, Caesarean section

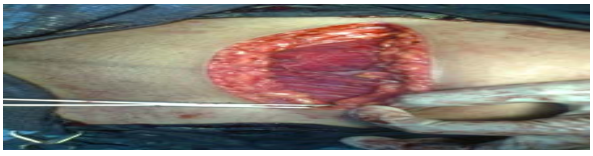
CASE REPORT :

A 21 year old primigravida with 36 completed weeks of pregnancy was referred to our institute from outside hospital for safe confinement and institutional delivery. Her antenatal period was uneventful. Since she had breech presentation obstetricians decided to deliver the baby by elective LSCS at 38 completed weeks. Preoperative evaluation : she was diagnosed to have glanzmann thrombaesthenia at the age of 9 years following frequent episodes of epistaxis and bleeding gums. platelet aggregation studies done previously revealed a positive response to ristocetin cofactor and no response to collagen, ADP, epinephrine. History of previous hospitalization for menorrhagia during which **she recieved whole blood and platelet transfusion** and was started on oral contraceptive pills. No history of previous surgeries or invasive procedures. **Haematologist opinion was obtained and was found to be variant type.** On examination: she had petechiae in upper limbs, B/L pitting pedal edema, no pallor, Ht:158cm, Wt:72kg, PR:86/min, BP:123/70mmHg, cardiovascular and respiratory system examination were normal.

Petechiae in left upper limb

Hematuria

Clear urine after platelet transfusion



Hemostasis achieved during surgery

Her blood investigation showed Hb-10.4gm/dl; PCV-31%; Platelet count-1.6lakh; Peripheral smearnormal platelet morphology**; Bleeding time-15min 30sec (nml-3 to 6min**); Clotting time-6min 30sec; Prothrombin time-12sec (control-13sec); INR-1.0(control-1.1); Activated partial thromboplastin time- 3osec(normal-27 to 35sec); renal and liver function tests were normal; blood grouping and typing-A positive; ECG-normal. Airway examination :MMS II; TMD and IID normal; neck movements and dentition normal; High risk informed consent was obtained. Blood bank informed about the need for blood and blood products. Patient was taken for surgery under ASA PS III On the morning of surgery, 4 units of platelets transfused,after which **bleeding time was found to be 3min 30 sec**. platelet transfusion was based on normalisation of bleeding time. inj.tranexemic acid 1gm and inj.calcium gluconate 10ml added to 500ml normal saline and infusion started at 125ml/hr. Inj.ranitidine 50mg and inj.metoclopramide10mg i.v given for acid aspiration prophylaxis.

Inside the operating room, monitors (ECG, NIBP, Pulse oximetry) connected. Bladder catheterized and urine was clear. Two intravenous lines with 16 G cannula secured in forearm veins. **Central neuraxial blockade was avoided because of the risk of spinal hematoma**. Premedicated with inj.glycopyrrolate

0.2mg i.v. preoxygenated with 100% oxygen for 5 minutes. Rapid sequence induction done with propofol 2mg/kg and succinylcholine 1.5mg/kg**. Gentle laryngoscopy** was done and intubated orotracheally with 7.0mm cuffed ETT. Anaesthesia was maintained with N2O:O2 (50:50) and sevoflurane 0.5-1% in a circle system. Atracurium 0.5mg/kg loading dose followed by 0.1mg/kg maintenance dose was used for muscle relaxation. Oxytocin 20 units infusion and inj.prostaglandin F2alpha 250micrg i.m was given after delivery of the baby. Intraoperative duration of surgery was 80min, with an estimated blood loss of 6ooml. Patient was haemodynamically stable throughout the procedure. Patient developed hematuria intraoperatively and **after 5 units of platelets transfusion** urine became clear. Haemostasis achieved**. Platelet transfusion was based on achieving hemostasis and normal bleeding time.** Residual neuromuscular blockade reversed with 10mkg/kg of glycopyrrolate and 50mkg/kg of neostigmine. After gentle and thorough oral suctioning, patients trachea was extubated. Postoperatively 4 units of platelets transfused as there was mild ooze from incision site. Inj.tranexemic acid 500mg 8th hourly continued for two days. No further ooze after platelet transfusion**.**

Bleeding time:6min 15sec; Ultrasonogram of abdomen revealed no free fluid in abdomen. Postoperative period was uneventful. She was discharged on 8th postoperative day with a healthy male baby.

DISCUSSION:

Glanzmann thrombaesthenia is an inherited bleeding disorder of megakaryocyte lineage characterized by lack of platelet aggregation due to deficient or dysfunctional GPIIb-IIIa receptors. Genes for both proteins are located on chromosome 17 and 50% activity of each protein is sufficient for normal platelet aggregation. Depending on the degree of receptor deficiency, fibrinogen binding, and clot retraction it can be classified into Type 1: <5% of normal glycoprotein 2b-3a receptor, absent fibrinogen binding and clot retraction. Type 2: 10-20% of normal glycoprotein receptors, moderately deficient clot retraction and fibrinogen binding Variant type: 50% of normal glycoprotein receptors, variant clot retraction and fibrinogen binding Platelet aggregation involves formation of GPIIb-IIIa- fibrinogen- GPIIb-IIIa complexes. This requires calcium and normal functioning of both glycoproteins. Clinical manifestations: epistaxis, gingival bleeding, excessive bleeding after dental extraction, petechiae, ecchymosis, menorrhagia, hemarthrosis, gastrointestinal haemorrhage and hematuria . Diagnosed by normal platelet count and morphology, prolonged bleeding time, decreased or absent clot retraction, platelet aggregation studies show absent response to platelet agonists :ADP, collagen, epinephrine and a normal response to ristocetin. In platelet function analyser, platelets fail to plug collagen based filter. Flow cytometry helps to detect glycoprotein deficiency. Clotting time, PT, APTT, INR will be normal. In this case, the disease was identified by platelet aggregation study.we used bleeding time to monitor the platelet function because other studies were not available in our hospital. Treatment of bleeding episode involves multiple platelet transfusions. The most common problem is development of antibodies to Human platelet antigen(HPA)1&2 present on GPIIb and GPa respectively. In patients with history of previous transfusions, presence of these antibodies must be ruled out using fluorescein conjugated monoclonal antibodies to GP2b/GP3a with flowcytometry. To avoid alloimmunization, l eukocyte depleted HLA matched platelets must be used. Gammaglobulin infusion can be used to diminish this antiplatelet response. Recombinant factor VII(rFVIIa) is an alternative to treat bleeding in patients with antiplatelet antibodies. This activates extrinsic pathway of coagulation in the absence of tissue thromboplastin and promotes formation of thrombin , which acts as a strong signal for recruitement of other platelets thereby forming tight fibrin plug. Dose of 90mkg/kg started two hours before surgery and repeated every 2hrly based on response. Desmopressin can also be used. It enhances platelet adhesion to vessel wall and shortens bleeding time. Topical agents like gelfoam and fibrin sealants are used to control bleeding. Antifibrinolytics eg.tranexemic acid can be used to prevent fibrinolysis.Drugs affecting platelet function must be avoided. (eg. NSAIDS, ticlopidine, clopidogrel, abciximab, dipyridamole.)Hepatitis B vaccination to prevent infectious risk associated with transfusion

Anaesthetic goals : Regarding anaesthetic management, Central neuraxial blockade must be avoided due to risk of spinal hematoma, Laryngoscopy and intubation must be gentle to avoid trauma to mucosa resulting in difficult airway due to bleeding, Invasive interventions like central venous cannulation must be done under ultrasound guidance to minimize trauma to tissues, Monitoring of platelet function is necessary(platelet aggregometry,bleeding time) to guide therapy, We must be prepared to manage severe bleeding which involves cooperation of blood bank staff and a multi therapeutic approach.

REFERENCES:

1) S.monte, G.lyons. **Peripartum management of a parturient with glanzmann thrombaesthenia** .Br J Anaesth 2002; 88: 734-38 .

2) Ercan xilmaz,Ugur Turhan. **Management of term pregnancy diagnosed with glanzmann thrombaesthenia: a case report.** Perinatal journal 2011; 19(3): 137-139.

3) Malhotra Neena, Chanana charu. **Pregnancy in a patient of glanzmann thrombaesthenia.** Indian journal of medical sciences. March 2006; 60(3):111-113.

4) Sherer D.M, Lerner R. **Glanzmann thrombaesthenia in pregnancy: a case and review of literature.** Am J Perinatology 1999; 16: 297-301.

5) Halil Ibrahim Uzunlar, Ahmet Eroglu. **Case report: A patient with glanzmann thrombaesthenia for emergency abdominal surgery.** Anaesth Analg Oct 2004; 99: 1258-60.