

A Rare Case Report on Pregnancy in Patient of Glanzmann's Thrombasthenia

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Abstract: *Glanzmann's thrombasthenia is a congenitally acquired platelet disorder with an autosomal recessive mode of inheritance. Though, quantitatively normal, the aggregation ability of platelets is reduced in this condition. Pregnancy and delivery are rare in these patients and have been associated with a high risk of severe postpartum haemorrhage. We describe a primigravida, who was diagnosed to have Glanzmann's thrombasthenia during adolescence. Patient was admitted with labor pain due to meconium - stained liquor after induction of labor emergency caesarean section was done, which was successfully managed by platelets and single donor platelet transfusion.*

1. Introduction

Glanzmann's thrombasthenia is a rare inherited disorder of platelet function transmitted as an autosomal recessive condition. The basic defect is the quantitative deficiency or a functional defect of the glycoprotein IIb - IIIa complex which results in bleeding diastasis due to defect in formation of a haemostatic plug. Pregnancy poses a special mention in these patients because of an increased risk of severe haemorrhage. We present the case of a primigravida who developed post - partum haemorrhage which was managed successfully with single donor platelet transfusion. Although various modalities have been used in the management of such patients there is no consensus. The use of single donor platelets seems to be beneficial as it not only raises the platelet counts but also the risk of antiplatelet antibodies is reduced.

2. Case Report

A 26 - year - old primigravida, a known case of Glanzmann's thrombasthenia presented to our labor room as emergency case with complaint of abdominal pain. The diagnosis of Glanzmann's thrombasthenia type II was made during a work up done for heavy and prolonged menstrual bleeding during adolescence at age of 15 year. However, at that time the bleeding was controlled with tranexamic acid and the patient required platelet transfusions. Patient had also history heavy menstruation during menses and which was every time controlled with tranexamic acid. She had also history of recurrent episodes of epistaxis.

The patient's pre pregnancy haematological work - up demonstrated haemoglobin of 11.4gm/dl, white cell count of 7600, platelet count 2.26 lacs, PT of 13 and APTT of 30.2 seconds. Bleeding time was >15 minutes. She had a spontaneous conception. All antenatal investigations including a detailed scan of the fetus was normal. Her antenatal course was uneventful except for two episodes of epistaxis during the second half of pregnancy at 26 and 31 weeks, which were managed conservatively. At term she was presented at labor room as emergency case with

complaint of abdominal pain with 9 month of amenorrhoea cephalic presentation with normal heart rate of fetus. On per speculum examination show present. On per vaginal examination cervical dilatation was 2 finger early effaced. Induction of labor planned. Induction done with dinoprostone PGE2. Continuous fetal heart rate monitoring done. All investigations were within normal limit Hb - 11.3gm/dl, platelet count was 2.56lacs. Bleeding time and clotting time was also within normal limit. After 4 hours of induction spontaneous ruptured of membrane occurred, meconium - stained liquor was present. Decision taken for emergency caesarean section. Pre operatively 2 unit of platelets were transfused patient shifted to operation theatre. Caesarean section was performed under general anaesthesia. Intra operatively 2 unit of platelets and 1 unit of single donor platelet were transfused. Healthy 3.2 - kilogram female baby was born. Estimated blood loss was approx one litter. Haemostasis achieved. Post - operative period was uneventful. Both the mother and baby were healthy and discharged 5 days after the caesarean section. Different contraceptive methods are advised.

3. Discussion

Glanzmann's Thrombasthenia is an inherited disorder of platelet function characterized by severe bleeding episodes. The laboratory studies show prolonged bleeding time with absent or decreased retraction and a normal platelet count. The coagulation studies are normal. Platelet aggregation in response to agonists ADP, collagen and arachidonic acid is absent. Clinical presentation of patients with the disorder includes haemorrhage symptoms like purpura, epistaxis, gingival haemorrhage and menorrhagia. These patients are at an increased risk of severe bleeding during pregnancy and in the intra and postpartum period. Although literature regarding pregnancy in patients of Glanzmann's thrombasthenia is limited, most authors have reported either peripartum or postpartum haemorrhage. An array of different modalities has been suggested for prevention and control of intra and postpartum haemorrhage in these patients.

In 1981, Sundquist et al, administered large doses of utero - tonics to prevent post - partum haemorrhage in their patient and were successful. Plasma - pheresis followed by platelet transfusions have been successfully used for prevention and treatment of intra and postpartum bleeding in cases of Glanzmann's disease. The rationale behind plasma exchange being to reduce the number of antiplatelet antibodies and hence making platelet transfusions hemostatically efficient.

The latest modality being used to correct postpartum haemorrhage in these patients is multiple doses of recombinant factor VIIa which is expensive. Pregnancy in patients with GT is rare, but it is life - threatening for both the patient and her fetus. The fetal risk is related to fetal immune thrombocytopenia induced by the transplacental passage of the maternal IgG antiGPIIb - IIIa isoantibodies, In case of the severe fetal thrombocytopenia, there is a risk of fetal intracranial haemorrhage. There is no standard of therapy or guideline regarding the management of GT with pregnancy nor any evidence is available because of uncommon incidence of this disease, rare possibility of this disease with pregnancy and ethical issue involved for any trial during pregnancy. The recombinant factor VIII though looks safe and efficient therapy, its multiple doses are required which is very expensive and will not be affordable by most of our patients.

Given this clinical scenario another option of "various normal platelet level with bleeding manifestations" validated in other situation may be followed. Maintaining normal platelets above 50, 000/mm³ LSCS and above 75, 000/mm³ for epidural anaesthesia is considered as safe limits for these procedures respectively. While planning platelet support in the setting, other factor to be considered is the median period of 5 days of platelet survival (external platelet support). The rate of rise of platelet count by one unit of S. D. P. /R. D. P. (40, 000/mm³ rise in case of one unit of S. D. P. Vs.6, 000/mm³ rise in one unit of / R. D. P.) and other diseases which may aggravate the bleeding severity. By applying the above principles of maintaining a safe and haemostatic platelet level, we could manage successfully high - risk pregnancy of GT.

Glanzmann's thrombasthenia during delivery. J Perinat Med.2004; 32: 456 - 8

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