PREGNANCY IN A PATIENT OF GLANZMANN'S THROMASTHENIA

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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 post-partum hemorrhage. We describe a primigravida, who was diagnosed to have Glanzmann's thrombasthenia during adolescence. She developed secondary post-partum hemorrhage after an elective caesarean section, which was successfully managed by single donor platelet transfusion.

Key words: Pregnancy, glanzmann's thrombasthenia, post-partum hemorrhage, single donor platelets

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 hemorrhage. We present the case of a primigravida who developed secondary post-partum hemorrhage 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.

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CASE REPORT

A 26-year-old primigravida, a known case of Glanzmann's thrombasthenia presented to our antenatal clinic in the first trimester of pregnancy. The diagnosis of Glanzmann's thrombasthenia type II was made during a work up done for heavy and prolonged menstrual bleeding during adolescence. However, the bleeding was controlled with tranexamic acid and the patient never required platelet transfusions. The patient's pre pregnancy hematological work-up demonstrated hemoglobin of 8.2gm%, hematocrit of 25.6%, white cell count of 7600, platelet count 2.26 lacs, PT of 13 and APTT of 30.2 seconds. Bleeding time was >15 minutes. No agglutination was noted with adenosine-di -phosphate, adrenaline, collagen and arachdonic acid in presence of ristocetin.

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 epistaxsis during the second half of pregnancy at 26 and 31 weeks, which were managed with nasal packing. At term she was planned for an elective caesarean in view of breech presentation. The hematologist planned transfusion of one unit of single donor platelets one hour prior to the surgery. Caesarean section was performed under general anesthesia and a healthy 3.2-kilogram female baby was born. Estimated blood loss was one liter. Post-operative period was uneventful. Both the mother and baby were discharged 5 days after the caesarean.

However, the patient returned to the hospital fifteen days later with a bout of heavy bleeding. Her vitals at this time were stable but her hemoglobin decreased to 5.2gm%. She was given intravenous oxytocin, prostaglandins and methylergometrine along with antibiotics but continued to bleed. She was transfused another unit of single donor platelets, after which her bleeding subsided. She remained in the hospital for 5 days following which she was discharged. The patient has been on regular follow-up since then and has not had any subsequent bout of bleeding.

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 arachdonic acid is absent. Clinical presentation of patients with the disorder includes hemorrhage symptoms like purpurae, epistaxsis, gingival hemorrhage 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 hemorrhage. An array of different modalities has been suggested for prevention and control of intra and postpartum hemorrhage in these patients.

In 1981, Sundquist et al,[1] administered large doses of utero- tonics to prevent post-partum hemorrhage in their patient and were successful. Plasma-pharesis followed by platelet transfusions have been successfully used for prevention and treatment of intra and postpartum bleeding in cases of Glanzmann's disease.[2] The rationale behind plasma exchange being to reduce the number of anti-platelet antibodies and hence making platelet transfusions hemostatically efficient. At our institution a case of Glanzmann's thrombasthenia who developed secondary postpartum bleeding was managed successfully with oral prednisolone.[3] There

is another case reported in literature where secondary post partum bleeding on the 14th day of delivery in a woman suffering from Glanzmann's disease was managed with oxytocin, prostaglandins, methylergometrine and tranexamic acid but bleeding abated only after platelet transfusion.[4] Sherer and Levner,[5] report a case in which the patient received 4 single donor platelet transfusions intrapartum, but she continued bleeding for 3 weeks post delivery for which she was transfused another unit of single donor platelets following, which the bleeding stopped. The latest modality being used to correct postpartum hemorrhage in these patients is recombinant factor VIIa.[6,7] Review of literature reveals lack of consensus regarding treatment of post-partum hemorrhage in patients with Glanzmann's thrombasthenia. Platelet transfusion is associated with a risk of isoantibody formation against glycoprotein IIb-IIIa.This results in a decreased efficacy of subsequent transfusions. Although the value of single donor platelet versus pooled platelet transfusion is not well established preparation of single donor platelets appears prudent and may assist in management of these patients who may have life threatening peripartum hemorrhage. Pregnancy in patient's with Glanzmann's thrombasthenia is rare, but the potential of post-partum hemorrhage should always be high in the minds of the managing obstetrician.

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