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Case Report

Pregnancy in two patients of Glanzmann's thrombasthenia: a rare case report

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ABSTRACT

Glanzmann's thrombasthenia (GT) is inherited 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 hemorrhage. We describe two GT here 1st case was a primigravida, who was diagnosed to have GT 11 yrs back and was admitted as a term pregnancy which was terminated by elective caesarean section and was successfully managed by platelet transfusion. 2nd case was a 24 year old lady got diagnosed as GT during the evaluation of frequent mucocutaneous bleed. Her antenatal management was like that of normal pregnancy. She was hospitalized 6 weeks prior to expected date of delivery. Elective LSCS (Lower Segment Caesarean Section) was done and was managed with 4 units of single donor platelet (SDP) without any other complication.

Keywords: Glanzmann's thrombasthenia, Gum bleeding, Pregnancy, Single donor platelet transfusion

INTRODUCTION

Glanzmann's thrombasthenia (GT) 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 due to defect in platelet aggregation and formation of a haemostatic plug.¹ Pregnancy poses a special mention in these patients because of an increased risk of severe hemorrhage². Although various modalities have been used in the management of such patients there is no consensus. We present two cases of primigravida with term pregnancy which was terminated by elective caesarean section and was successfully managed by platelet transfusion.

CASE REPORT

Case 1

A 36-year-old elderly primigravida, a known case of GT presented to our antenatal clinic at 36 wks for confinement. The diagnosis of GT was made 11 yrs back at clinical Haematology Department of our institution when she was admitted as a case of menorrhagia with frequent gum bleeding, not adequately responding to medications for which she was evaluated with tests for coagulation defects, platelet functions and for thrombosis besides other routine haematological investigations. On evaluation platelet count was normal with no platelet aggregation with adenosine-di-phosphate, adrenaline, collagen and arachdonic acid but normal aggrergation in

presence of ristocetin, flow cytometric analysis revealed CD 41 (Gp IIb) and CD 61 (Gp IIIa) expression <1%. The coagulation screening tests like prothrombin time (PT), activated partial thromboplastin time (aPTT), serum fibrinogen and fibrinogen degradation products were within normal limits. The Vonwillbrand factor assay, liver function tests, blood urea, serum creatine and other biochemical parameters were within normal limits. She was diagnosed to be a case of GT. She was satisfactorily managed with tranexamic acid for frequent gum bleeding, petechial spots, menorrhagic cycles, prolonged bleeding on minor trauma without any platelet transfusion. She had no history of consanguineous marriage in her family and none of her first degree relatives had any bleeding or clotting disorders. During her follow up in haematology department both wife and husband expressed their desire to have pregnancy. They were counseled regarding the risks involved, the possibility of various complications and the need for hospitalization and pregnancy management in a tertiary care centre with availability of all required facilities along with platelet support. They agreed on the above points and planned for pregnancy. The patient conceived on 14 September 2014 and referred to our antenatal clinic.

During her 1st antenatal visit to our antenatal clinic patient's hematological work-up demonstrated hemoglobin of 6.6 gm%, hematocrit of 22%, white cell count of 7900, platelet count 2.4 lacs, PT of 10.7 seconds (Normal value within 3-5 sec of control) and APTT of 21.7 seconds (Normal value within 5-7 sec of control, INR 0.92, Bleeding time was 1 mins 50 seconds (Normal value 3-5 min), Clotting time 4 mins 15 sec (Normal value 3-12 min. Comment on peripheral smear revealed microcytic hypochromic blood picture. Fibrin degradation product was negative, low (10.6ng/ml) (Normal value up to 200ng/ml) and normal Ferritin levels, Direct Coombs Test negative. One of her incidental finding was S.TSH 6.13. As per the advice of haematologist, she was transfused with whole blood daily for three days and Tablet Thyroxine 75 microgram was started as per the advice of endocrinologist. She was advised to continue ferrous salt at the dose of 2mg /kg orally along with folic acid 5 mg/day. Her subsequent visits were uneventful and advised for regular antenatal check-up, to intake iron and calcium regularly and admitted to antenatal ward at 36 weeks.

On basis of non-reassuring CTG and low-lying placenta she was planned for elective LSCS. As per haematologist advice she was transfused with one unit of single donor platelets one day before surgery, another unit one hour prior to the surgery. Caesarean section was performed under general anesthesia and a healthy 2.3 kilogram female baby was born. Estimated blood loss was one liter, there was oozing from the stitch site which was managed by diathermy and injection tranexamic acid. Patient could not be extubated after surgery and required ventilatory support for one day, during which she was transfused with two units of fresh blood and six units of

random donor platelets, after which she improved. She again received one unit of single donor platelet on third postoperative day. CS scar site was healthy, her non-absorbable skin stitches were removed on sixth postoperative day. She received four units of random donor platelets daily for three consecutive days starting from eighth post-op day. Mother with baby was discharged on 11th post-operative day with normal haematological investigation reports. The patient has been on regular follow-up since then and has not had any subsequent bout of bleeding.

Case 2

One 24 year old lady from western part of Odisha got diagnosed as GT during the evaluation of frequent mucocutaneous bleed. She was requiring tranexamic acid and platelet support intermittently for menorrhagia. She got marriage and after 2 years expressed her desire for pregnancy. Both wife and husband were counseled like that of first case and referred to our antenatal clinic after pregnancy. Her antenatal management was like that of normal pregnancy. She was hospitalized 6 weeks prior to expected date of delivery. Elective LSCS was done and was managed with 4 units of single donor platelet (SDP) without any other complication.

DISCUSSION

Glanzmann's Thrombasthenia (GT) 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 hemorrhage symptoms like purpurae, epistaxis, gingival hemorrhage and menorrhagia. These patients are at an increased risk of severe bleeding during pregnancy and in the intrapartum and postpartum period.^{2,3} Although literature regarding pregnancy in patients of GT 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 administered large doses of uterotonics to prevent post-partum hemorrhage in their patient and were successful.⁷ Plasmapheresis followed by platelet transfusions have been successfully used for prevention and treatment of intrapartum and postpartum bleeding in cases of Glanzmann's disease.⁸ The rationale behind plasma exchange is to reduce the number of anti-platelet antibodies and for making platelet transfusions hemostatically efficient. Cases of Glanzmann's thrombasthenia have been reported who developed secondary postpartum bleeding and managed successfully with oral prednisolone.⁹ 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.¹⁰ Sherer and Levner, 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.¹¹ The latest modality being used to correct postpartum hemorrhage in these patients is multiple doses of recombinant factor VIIa which is expensive.^{12,13}

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 anti-GPIIb-IIIa isoantibodies. In case of the severe fetal thrombocytopenia, there is a risk of fetal intracranial hemorrhage.

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 look 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 are 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 these two cases of high risk pregnancy of GT. Blood components along with platelet (S.D.P./R.D.P.) are now available in many centers and also affordable. But this high pregnancy should be managed preferably in a tertiary care centre where interdepartmental facilities and co-operation among obstetricians, haematologists, transfusion physicians etc. are available.

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