

Glanzmann's Thrombasthenia and Pregnancy

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Abstract

Glanzmann's thrombasthenia is rare autosomal recessive platelet disorder caused by abnormal platelet glycoprotein complex (GP IIb-IIIa) leading to excessive mucosal bleeding. Patients with Glanzmann's thrombasthenia may present with hemorrhagic symptoms including mucosal bleeding, easy bruising and post-surgical bleeding. We report a 37-year old lady, who underwent a cesarean section at term. She received recombinant factor VIIa at the time of surgery. She had uneventful post operative period and she was discharged home in good general conditions.

Keywords: Glanzmann's thrombasthenia, pregnancy, bleeding.

(J Med J 2009; Vol. 43 (2):134-136)

Received

Accepted

May 18, 2008

September 25, 2008

Introduction

Glanzmann's thrombasthenia is an inherited hemorrhagic disorder characterized by severe reduction or absence of platelet aggregation due to qualitative or quantitative abnormalities of platelet glycoprotein IIb-IIIa.¹ Pregnancy is rare in patients with Glanzmann's thrombasthenia. Pregnant patient may develop bleeding during pregnancy due to gynecologic or obstetric causes and bleeding may occur during and after delivery² and even during puerperium.³ Treatment of bleeding episodes may require platelet transfusion. However, repeated platelet transfusion may result in antiplatelet antibodies formation. Recombinant factor VIIa has been shown to be effective in treating severe bleeding resulting in Glanzmann's thrombasthenia.⁴ We report a 37-year old lady, who underwent a cesarean section at term. She received recombinant factor VIIa at the time of surgery.

She had uneventful postoperative period and she was discharged home in good general conditions with uneventful postpartum period.

Case Report

A 37-year-old lady was diagnosed to have Glanzmann's thrombasthenia with antibodies to GP IIb-IIIa when she was 6 years old. She presented for the first time to our clinic 4 years back complaining of primary infertility for a duration of 6 years duration. She gave history of many admissions to different hospitals complaining of bleeding including epistaxis and intraabdominal bleeding resulting from ovulatory follicular rupture. She was managed by blood and platelet transfusion. She was investigated thoroughly and she was diagnosed to have unexplained infertility and she was advised to undergo in vitro fertilization (IVF).

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After 3 years she presented with missed period and she was diagnosed to have missed abortion at 9 weeks gestation. She underwent suction evacuation after she received one unit of single donor platelet. Her postoperative period was uneventful. After 3 months she presented with missed period and she was diagnosed to be pregnant she had uneventful antepartum period with no obstetric complications. She was planned for an elective cesarean section at term due to breech presentation. Hematology plan was to prepare recombinant factor VIIa, four units of filtered RBCs and one unit of single donor platelet. At the day of surgery after induction of anaesthesia and with skin incision, the patient received recombination factor VIIa at the dose of 150 µg/kg, three vials push over 3 minutes and was continued according to table 1. The patient underwent smooth cesarean section and the outcome was an alive female baby weighing 3.1 kg with an apgar score of 7/9 and went to normal nursery. After closure of the uterus, the patient started to have oozing, where she received another 3 vials of a recombinant factor VIIa and bleeding stopped. She received a total of 24 vials of recombinant a factor VIIa according to the schedule (Table 1) and last one was on the third day postpartum, where she was discharged in good general conditions. She was seen one week and one month later in outpatient clinic. She was in good general conditions and she had no complaints.

Table (1): The schedule of recombinant factor VIIa received by the patient.

<i>Time</i>	<i>Dose</i>
<i>0h(skin incision)</i>	<i>3 vials I.V. push (over 2-3m)</i>
<i>3h</i>	<i>3 vials I.V. push</i>
<i>9h</i>	<i>3 vials I.V. push</i>
<i>17h</i>	<i>2vials I.V. push</i>
<i>29h</i>	<i>2vials I.V. push</i>
<i>Q12h</i>	<i>2vials I.V. push</i>

Discussion

Patients with Glanzmann's thrombasthenia may develop mucosal bleeding including epistaxis, skin bruises and they may develop severe bleeding after any surgical procedure.

Gynecologists are faced with these patients when they develop menorrhagia during menses or with intraabdominal bleeding at the time of ovulation because of ruptured follicle during ovulation. Furthermore, they may present with hemorrhagic ovarian cyst. Once pregnancy occurs, patients may develop bleeding during pregnancy for obstetric causes, intrapartum or postpartum.

Pregnancy in patients with Glanzmann's thrombasthenia is rare,⁵ but it is life-threatening for both the patient and her fetus.⁶ 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 lack of consensus regarding treatment of postpartum hemorrhage in patients with Glanzmann's thrombasthenia. The presence of isoantibodies to GPIIb-IIIa may be harmful for both the mother and her fetus. To prevent the platelet immunization, platelet transfusion should be limited and recombinant factor VIIa is an alternative. Treatment options in this type of patients is limited to a single dose of platelet transfusion or/ and to the recombinant factor VIIa. A team work, the obstetrician and the hematologist is essential in deciding the best option and best time to interfere in this high- risk group of patients.

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