



Management of Term Pregnancy Diagnosed with Glanzmann Thrombasthenia: a Case Report

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Abstract

Objective: We aimed to discuss the case with Glanzmann thrombasthenia and caesarean performed.

Case: Twenty-one years old patient with a diagnosis of Glanzmann thrombasthenia was delivered by cesarean section.

Conclusion: Glanzmann thrombasthenia had been diagnosed in early childhood and including mucocutaneous bleeding, epistaxis and menorrhagia. The lack of platelet aggregation in Glanzmann thrombasthenia leads to mucocutaneous bleeding whose manifestation may be clinically variable, ranging from easy bruising to severe and potentially life-threatening hemorrhages.

Keywords: Thrombocyte adhesion, delivery, obstetric, pregnancy, trombasthenia.

Glanzmann trombastenisi tanısı alan term gebenin yönetimi: Olgu sunumu

Amaç: Glanzmann trombastenisi tanısı alan ve sezaryen ile doğum yaptırılan olguyu tartışmayı amaçladık.

Olgu: Yirmibir yaşında Glanzmann trombastenisi tanısı alan hasta sezaryen ile doğurtuldu.

Sonuç: Glanzmann trombastenisi erken çocukluktan itibaren mukokutanöz kanama, epistaksi ve menoraji olarak kendini gösterir. Glanzmann trombastenili hastalarda trombosit adezyonundaki bozukluk nedeniyle ciddi hemorajiye neden olan mukokutanöz kanamalarla kendini gösterir.

Anahtar Sözcükler: Trombosit adezyonu, gebelik, doğum, trombasteni.

Introduction

Glanzmann's thrombasthenia (GT) is an uncommon autosomal recessive disorder characterized by lack of platelet aggregation despite of normal platelet count and morphology.^[1-5] There are qualitative and quantitative abnormalities in glycoprotein IIb/IIIa (GP integrin proteins which work as a receptor for fibrinogen and von Willebrand factor (vWF) on the platelet surface.^[2] More than fifty mutations have been identified in these GPs.^[6] Epistaxis, mucocutaneous hemorrhage, bleeding gums and menorrhagia in girls are the most common symptoms of the early childhood. Laboratory studies show that prolonged bleeding time, normal or subnormal platelet count and clustered platelets in peripheral smear. Platelets exhibit aggregation in the presence of ristocetin. GT diagnosis should

confirmed by flow cytometry. Currently approach to Glanzmann thrombasthenia; flow cytometry and glycoprotein analyses are excellent. Flow cytometry is particularly beneficial for the diagnosis of aGT, being the only test able to characterize both the functional effect and the molecular target of the patient's autoantibody. There are three subtypes for GT. In type 1 GP IIa/IIIb expression is smaller than 5% and causes more severe bleeding because of alpha granule fibrinogen deficiency and coagulation reaction failure. Type 2 is a less severe form in which GP IIa/IIIb expression is about 10-20% leading to difficulties in coagulation. Type 3 mostly results from qualitative GP abnormalities and has a GP expression greater than 50%. Globally, it is extremely rare but it has a relatively high incidence in consanguineous populations where intermarriage is common.^[7]

Case Report

A 21 years old who had diagnosed Glanzmann's thrombasthenia during her childhood was admitted to our obstetrics clinic. Her previous pregnancy had been ended by elective C/S because of breech presentation in Malatya State Community Hospital where she was also given activated factor VII derivatives (Novoseven, 90 mcg/kg, preoperative two days, twice a day) and four units of fresh frozen plasma(FFP) and two units of whole blood intra operatively. In the history of the patient, admitted to the hematology clinic due to heavy menstrual and nasal bleeding when she was 15 years old. Physical examination has been revealed no abnormalities. Hemoglobin levels were 7.6 g/dl. Platelet count, prothrombin time (PT), and partial thromboplastin time (PTT) were normal. The bleeding time performed by the Ivy method was 16 min. The patient received 6 U of packed red blood cells. Treatment with conjugated estrogen tablets oral Tranexamic acid 500 mg three times daily was instituted. Obstetric ultrasonography (USG) was compatible with 38 week gestational age and blood evaluations were as follows. Hemoglobin (hb): 11.9 mg/dl, bleeding time 13.4 minute. White blood cell count (WBC) 11.300 cells/IL/cu mm, platelet count (PLT): 156,000/ml. Activated partial thromboplastin time (APTT): 30.7 second, International Normalized Ratio (INR): 1.1, fibrinogen: 213,000 g/L. She was scheduled for a C/S with previous C/S and Glanzmann's Trombasthenia diagnosis. She was given one unit of manual thrombocyte and undergone an operation. A rapid sequence induction of general anesthesia was performed after preoxygenation, using thiopental 6 mg/kg and succinylcholine 1.5 mg/kg, followed by oral intubation of the trachea. Skin was passed by Pfannenstiell incision and the uterus was cut by Kehr incision. 3,300 grams healthy male baby (whose PLT was 290,000) was born. He was also assessed in terms of intracranial hemorrhage by pediatricians. Since blood leakage from uterine Kehr incision line and other peritoneal surfaces was observed, two drainage tubes intraabdominally and one subcutaneously was placed intraoperatively. Estimated blood loss was about 1,000- 1,100 cc. First intraoperative laboratory result for hb and PLT were 7.3 g/dL and 209,000/mL respectively. After two units of erythrocyte suspension administration her control hb value 8.4 gr/dL. Other result were as following PLT: 128,000/mL, APTT: 26.4 second, INR: 1.2, fibrinogen: 198,000 g/L. After first control evaluations she was replaced to her room. In the first 24 hours under clinic observation, her vitals were stable. She was given 15 units of manual tromboocyte. 350 cc sero-hemorrhagic liquid was collect-

ed in the abdomen drains. Her ecchymotic skin areas around the incision were treated by mucopolysaccharide polysulphate containing pomade. In the second postoperative 24 hours her vitals were stable and hb value was 9.6 g/dL. 5 cc and 25 cc serohemorrhagic liquids were collected in subcutaneous and abdominal drains respectively. In the day 3 postoperatively, drainage tubes were removed on the account of any drainage. Her blood values were followed closely for four more days and she was discharged with her healthy baby on the eighth day postoperatively.

Discussion

Our knowledge about Glanzmann's thrombasthenia is restricted today. Thrombocytopenia is seen in 4% of all pregnancies. Platelets counts are not below 50,000/L urgent obstetric precautions are not needed. Patients should be differentiated from ITP which commonly can be seen, gestational thrombocytopenia and especially Hellp syndrome. Physicians should also keep in mind that gestational thrombocytopenia which doesn't have a special treatment is only seen in the last trimester of pregnancy.

The patients with thrombocytopenia diagnosis should be considered accurately in terms of perinatal and postnatal hemorrhage and treatment should be applied with the suggestions of Hematology. In 1981, risk of peripartum bleeding tended to be decreased by use of uterotonic agents at overdose.^[8,9] For such patients, transfusion of platelets in preoperative and postoperative periods was successful. According to another GT patient case report with excessive vaginal bleeding at the 14th day of C/S, prostaglandin, methergine, oxytocin, methylergometrine and tranexamic acid attempts was found to be meaningless.

After administration of platelet transfusion in this patient, diminished bleeding was also reported.^[10] Desmopressin (dDAVP) has been attempted as therapy but has not shown any proven clinical usefulness.^[11] Bone marrow transplants have been used successfully in rare cases, though this remains a drastic treatment.^[12,13] In addition, desmopressin acetate (DDAVP) has been shown to be effective in preventing the bleeding in some of the platelet dysfunction syndromes.^[14] However, as the platelet transfusions given preoperatively can contribute hemostasis, occasionally it may also cause formation of antibodies against platelets.^[15] Plasma Exchange decreases these platelet antibody formations.^[6] Expensive activated factor VII derivative is the effective cure way. It is activated by tissue factor released from damaged tissue and leads to fac-

tor X activation at the injury site. It is well known that in liver diseases synthesis of factors also decreases significantly.^[15]

In 1996, Terngborn et al. defined that use of activated factor VII in persistent epistaxis was essential.^[16] Later from that plenty of published data manifested its beneficial effects.^[17] We cured that patient with factor VIIa even though he had an intact hemostatic system. The development of this coagulation factor is a recent addition to the available treatments for patients with hemophilia and inhibitors to factor VIII or IX.^[18] It has also been used in patients with thrombocytopenia, congenital defects in the platelet glycoproteins,^[19] and acquired thrombocytopathia.

Recombinant activated factor VII (rFVIIa; NovoSevenP, Novo Nordisk, Denmark) triggers hemostasis heavy threatening bleeds and in major surgery in hemophilia A and B patients and also more than 180,000 standard doses of rFVIIa had been administered from 2001.^[20] rFVIIa induces hemostasis by enhancing thrombin generation on the thrombin-activated platelet surface, thereby providing the formation of a fully stabilized, tight fibrin hemostatic plug, which is resistant to premature fibrinolysis. rFVIIa is an excellent and attractive alternative to platelet transfusions for the treatment of dysfunctional platelet, attached bleeding. Nevertheless, this needs to be corroborated on a larger population.

We must have experience with other studies to determine the minimal effective dose, the ideal way to administer rFVIIa, the relative role of rFVIIa and antifibrinolytic agents in these patients who admit heavy bleeding, as well as the prime cost and efficacy of using rFVIIa.

Conclusion

Glanzmann thrombasthenia is diagnosed in childhood and it is a bleeding disorder that may cause severe bleeding diathesis. However, surgery can be successful if the necessary precautions taken. Pregnancies of patients diagnosed in childhood have not received antenatal follow-up during the birth, as well as the disease causes severe anemia or bleeding disorders. As in this case report of antenatal follow-up of patients diagnosed in childhood, and any complications during the birth necessary precautions are taken.

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