Magudapathi et al., J Clin Case Rep 2012, 2:8
DOI: 10.4172/2165-7920.1000142

Journal of Clinical Case Reports

Case Report Open Access

Glanzmann's Thrombasthenia Complicating Pregnancy

Chandrakala Magudapathi[1]*, Suthanthira Kannan[2] and Deepa Giri[3]

[1]Consultant, Department of Obstetrics & Gynaecology, G. Kuppuswamy Naidu Memorial Hospital, Nethaji road, Coimbatore, India
[2]Consultant, Department of Haemato oncology, G. Kuppuswamy Naidu Memorial Hospital, Nethaji road, Coimbatore, India
[3]Registrar, Department of Obstetrics & Gynaecology, G. Kuppuswamy Naidu Memorial Hospital, Nethaji road, Coimbatore, India

Abstract

Glanzmann's thrombasthenia is a rare autosomal recessive hemorrhagic disorder caused by abnormal platelet glycoprotein complex (GP IIb-IIIa) presenting with hemorrhagic symptoms. Pregnancy is not uncommon because fertility is not affected but an association is rare. Also delivery often results in haemorrhage. Newborn thrombocytopenia is occasionally severe, but is always transitory. We report a 21-year old primigravida, who underwent vaginal delivery at term with IUGR and Oligohydramnios. Glannzmann's thrombasthenia was characterized by aggregation and this patient went undiagnosed till adolescence until evaluated for puberty menorrhagia which is uncommon. She received single donor platelet transfusion during the intrapartum period along with tranexemic acid. Platelet transfusion may result in alloimmunization and make successive transfusions less effective. We did not use Factor VII in the intrapartum period due to cost and suspected fetal effects. Postnatally she had hematuria settled on the 2nd day with conservative management. We present this case because of association of Glanzmann's thrombasthenia with IUGR and oligoamnios the only reported case so far.

Keywords: Glanzmann's thrombasthenia; Pregnancy; IUGR

Introduction

Glanzmann's thrombasthenia is an autosomal recessive hemorrhagic disorder characterized by a severe reduction in or absence of platelet aggregation in response to multiple physiologic agonists. It is due to qualitative or quantitative abnormalities of platelet receptor GPIIb/IIIa required for platelet aggregation. Pregnancy poses a special mention in these patients because of an increased risk of severe hemorrhage.

Case Report

A 21-year-old primigravida with Glanzmann's thrombasthenia presented to our antenatal clinic in the second trimester of pregnancy. The diagnosis of Glanzmann's thrombasthenia was made during a work up done for puberty menorrhagia. The bleeding was controlled with tranexamic acid and progesterone. She never required platelet transfusions. She had history of prolonged bleeding following injections or injuries during childhood. The patient's pregnancy work-up was showing hemoglobin of 9.5 gm%, white cell count of 13,000/mm, platelet count 2.5 lakhs, a PPT of 25/29. Bleeding time was >10 minutes and clot retraction time absent.

She had a spontaneous conception as fertility is usually not compromised in these patients. All antenatal investigations including a detailed scan of the fetus were normal. Her antenatal course was uneventful in the first and second trimesters except for moderate anemia for which she was on oral hematinics. She was diagnosed with IUGR and Oligohydramnios AFI 5 cm at 37 weeks and induced with prostaglandins and monitored closely. She progressed to deliver vaginally a 2.240 kilogram female baby. The first stage lasted for 7 hours 15 min, the second stage lasted for 25 min and 2 units of platelet-rich plasma were infused. Episiotomy was performed followed by instrumental delivery due to fetal bradycardia (6min). There was no additional bruising or laceration of the perineum. The third stage was uneventful. Estimated blood loss was 250ml. An oxytocin infusion of 20 units in 500 ml of ringer lactate was started immediately following delivery of the anterior shoulder.

Patient had hematuria which settled on the second post natal day. Baby was in NICU for respiratory distress and phototherapy in view of hyperbilirubinemia- which on evaluation was found to be physiological and not a manifestation of Glanzmann's thrombasthenia. Patient's haemoglobin after 48 hours was 8.2gm%. Mother and baby were discharged on third post natal day in good condition. During her follow up visit 15 days later patient had heavy bleeding for which she was treated with tranexemic acid and it settled in 2 days.

Discussion

Glanzmann's Thrombasthenia is a rare inherited bleeding disorder caused by a deficiency or dysfunction of the GPIIb-IIIa receptor on platelets. Glanzmann, a Swiss pediatrician, initially described thrombasthenia in 1918 when he noted purpuric bleeding in patients with normal platelet counts [1].

Over 500 cases of thrombasthenia have been reported in the international literature. Although GT predominates among certain ethnic groups (Arab populations, Jordanian nomadic tribes, Iraqi Jews, French gypsies, and individuals from southern India), an estimate of worldwide incidence and prevalence has not been reported. The incidence is more in families with consanguinity [2].

Death following bleeding is estimated at approximately 5-10% mostly due to occurrence of severe unprovoked intracranial or GI hemorrhages.

The gene for GPIIb –IIIa is carried on chromosome 17 in humans so it affects men and women equally. Many patients with identified mutations are compound heterozygotes being higher where consanguinity is common. At the present time, 38 mutations in GPIIb

***Corresponding author:** Chandrakala Magudapathi, Consultant, Department of Obstetrics & Gynaecology, G. Kuppuswamy Naidu Memorial Hospital, Nethaji road, Coimbatore, India, E-mail: chandrakala_m@yahoo.com

Received April 10, 2012; **Accepted** April 30, 2012; **Published** May 14, 2012

Citation: Chandrakala M, Suthanthira K, Deepa G (2012) Glanzmann's Thrombasthenia Complicating Pregnancy. J Clin Case Rep 2:142. doi:10.4172/2165-7920.1000142

and 25 mutations in GPIIIa have been recorded. It is speculated that a 15; 17 translocation characteristic of acute promyelocytic leukemia may interfere with GPIIb and GPIIIa genes, located at 17q21.32 [3].

The molecular characterization of Glanzmann's thrombasthenia in patients and their families has permitted DNA-based carrier detection and prenatal diagnoses to be performed [4]. Acquired thrombasthenia due to glycoprotein IIb-IIIa platelet antibodies has also been defined in several conditions [5].

Patients with GT are typically diagnosed in infancy or early childhood. However, it can also be diagnosed in adult. The most common clinical manifestations of Glanzmann's thrombasthenia are menorrhagia, easy bruising, purpura, epistaxis and gingival bleeding. Less common are gastrointestinal hemorrhage and hematuria. Hemarthrosis and intracranial hemorrhage are rare. Carriers of Glanzmann thrombasthenia appear to be asymptomatic [6].

Glanzmann's thrombasthenia is classified into three types, depending on the level of GPIIb-IIIa present. The clinical manifestation does not depend on the severity.

Type 1 (severe) : <5% of normal GPIIb-IIIa levels

Type 2 (less severe): 10-20% of normal GPIIb-IIIa levels.

Type 3 (variant): normal levels of GPIIb-IIIa, but functionally inactive.

The goal of treatment is to control bleeding episodes.

Platelet transfusion is the standard therapy. However, approximately 15-30% of patients become refractory to platelet transfusion or develops antibodies to GPIIb-IIIa and/or HLA antibodies.

Factor VII is indicated for the treatment of bleeding episodes and for the prevention of bleeding during surgery or invasive procedures in patients with Glanzmann's thrombasthenia with antibodies to GPIIb-IIIa and/or HLA, and with past or present refractoriness to platelet transfusions.

Desmopressin (DDAVP) has been tried in some patients with Glanzmann's Thrombasthenia and may shorten bleeding time in patients with type 2 only, but there is no notable clinical efficacy.

Oral contraceptives can regularise menstrual cycles and reduce the bleeding. This is sometimes recommended before a girl's first period, as haemorrhage is particularly severe at this time.

Immunoabsorption is the removal of antibodies to platelets by plasma exchange with the use of protein-A sepharose columns which may transiently restore platelet efficacy. However this technique is not available everywhere, it is labour intensive and requires an adequate venous access. It is not effective in active bleeding as these process reqs.

Allogeneic marrow transplant has been reported in two patients with Glanzmann's thrombasthenia.

Other Treatments

Compression, gelatine sponge or gauze, antifibrinolytic agents such as tranexamic acid or topical thrombin can be used to control minor bleeds.

Conclusion

Pregnancy is rare in patients with Glanzmann's thrombasthenia [7] and pregnancy is life threatening for both mother and fetus [8]. Pregnant patient may develop bleeding due to gynecologic or obstetric causes and bleeding may occur during and after delivery [9] and even during puerperium [10]. There is lack of consensus regarding treatment of postpartum hemorrhage in patients with Glanzmann's thrombasthenia. Studies show that large doses of utero- tonics prevent post-partum hemorrhage in the patient [11]. Plasmapheresis followed by platelet transfusions have also been successfully used for prevention and treatment of intra and postpartum bleeding in cases of Glanzmann's disease as this reduces the number of anti-platelet antibodies making transfusions effective [12]. The latest modality being used to correct postpartum hemorrhage in these patients is recombinant factor VIIa [13,14]. Oral prednisolone has also been used to treat secondary PPH in some centres [15].

References

1. Glanzmann E (1918) Hereditare hamorrhagische thrombasthenie. Ein Beitrag zur Pathologie der Blutplattchen. J Kinderkranken 88: 113.
2. Nurden AT (2006) Glanzmann thrombasthenia. Orphanet J Rare Dis 1: 10.
3. Bray PF, Barsh G, Rosa JP, Luo XY, Magenis E, et al. (1988) Physical linkage of the genes for platelet membrane glycoproteins IIb and IIIa. Proc Natl Acad Sci U S A 85: 8683-8687.
4. French L, Seligsohn U (2000) Platelet glycoprotein IIb/IIIa receptors and glanzmann's thrombasthenia. Arterioscler Thromb Vasc Biol 20: 607-610.
5. Bloor AJC, Smith GA, Jaswon M, Norman EP, Willem HO, Ri L (2006) Acquired thrombasthenia due to GPIIbIIIa platelet autoantibodies in a 4-year-old child. Eur J Haematol 76: 89-90.
6. George JN, Caen JP, Nurden AT (1990) Glanzmann's thrombasthenia: the spectrum of clinical disease. Blood 75: 1383-1395.
7. Malhotra N, Chanana C, Deka D (2006) Pregnancy in a patient of Glanzmann's thrombasthenia. Indian J Med Sci 60: 111-113.
8. Leticee N, Kaplan C, Lemery D (2005) Pregnancy in mother with Glanzmann's thrombasthenia and isoantibody against GPIIb-IIIa: Is there a foetal risk? Euro J Obstet Gynecol Reprod Bio 121:139-142.
9. Sherer DM, Lerner R (1999) Glanzmann's thrombasthenia in pregnancy: a case and review of the literature.Am J Perinatol 16: 297-301.
10. Capuzzo E, Polatti F, Zara C (1997) Glanzmann's thrombasthenia and puereperium. Int J Gynecol Obstet 57: 313-314.
11. Sundqvist SB, Nilsson IM, Svanberg L, Cronberg S (1981) Pregnancy and parturition in a patient with severe Glanzmann's thrombasthenia. Scand J Haematol 27: 159-164.
12. Ito K, Yoshida H, Hatoyama H, Matsumoto H, Ban C, et al. (1991) Antibody removal therapy used successfully at delivery of a pregnant patient with Glanzmann's thrombasthenia and multiple anti-platelet antibodies. Vox Sang 61: 40-46.
13. Kale A, Bayhan G, Yalinkaya A, Yayla M (2004) The use of recombinant factor VIIa in a primigravida with Glanzmann's thrombasthenia during delivery. J Perinat Med 32: 456-458.
14. Shamsi TS, Hossain N, Soomro N, Hasan JA, Noorani M et al. (2005) Use of recombinant factor VIIa for massive postpartum haemhorrage: case series and review of literature. J Pak Med Assoc 55: 512-515.
15. Kashyap R, Kriplani A, Saxena R, Takkar D, Choudhry VP (1997) Pregnancy in a Patient of Glanzmann's Thrombasthenia with antiplatelet antibodies. J Obstet Gynaecol Res 23: 247-250.