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Case Report

A case of Glanzmann's thrombasthenia type I in a primigravida treated with factor VIIa

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Glanzmann's thrombasthenia is a rare autosomal recessive platelet function disorder. Though rare in the global context, this disorder is relatively more common in communities where consanguineous marriages are more frequent. The defect in platelet function is the inability of thrombasthenic platelets to aggregate. Their structural defect is the deficiency or dysfunction of platelet membrane (glycoprotein) GpIIb/IIIa complex which mediates platelet aggregation via fibrinogen binding. Epistaxis, gum bleeding, menorrhagia are the common clinical manifestations, whereas large muscle hematoma or hemarthrosis seldom occur in these patients. They often suffer from severe bleeding complications in case of injury or surgery. Pregnancy is uncommon and dangerous

Case report

Mrs. SJ, a 24 year old primigravida presented at our casualty on 13th June 2005 complaining of pain in the abdomen. Her expected date of confinement was 20th June 2005. She was on regular antenatal check up and her antenatal period was uneventful. She gave a history of increased bleeding tendencies since 2 years of age. Initially she had prolonged bleeding from the gums. Subsequently she had episodes of post-traumatic, superficial, subcutaneous hematoma, gastrointestinal bleeding, mildly prolonged bleeding after trauma and two episodes of epistaxis and purpura. For these symptoms she had been over a period of time, transfused more than 150 units of fresh whole blood and blood products. She was born to second degree consanguineous parents and there was no definite family history of similar illness.

At the Christian Medical College at Vellore she was diagnosed to have Glanzmann's Thrombasthenia Type 1 (severe Type). This was based on clinical history and laboratory results which showed a normal platelet count and morphology, normal clotting time, and abnormally prolonged bleeding time, and no clot retraction indicating a primary hemostatic failure of a platelet nature. To confirm an aggregometer study revealed absence of aggregating response with agonists ADP, adenosine diphosphate, epinephrine, collagen but a normal response with ristocetin indicating the absence of platelet surface glycoprotein receptor Gp 11b/111a.

Her investigation reports were as follows: hemoglobin 7.59g/dL, packed cell volume 25%, platelet count 2.1 lakh, bleeding time 50 minutes, clot retraction nil. She

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was given one unit of platelet concentrate and one unit of packed cells. On June 20th 2005 labor was induced with PGE 2 gel for nonreactive nonstress test after giving one unit of single donor platelets. A decision to do LSCS was made on June 21st for non progress of labor. A female baby weighing 3.7 kg was delivered by FT LSCS. During surgery bleeding was more than normal and she was given one unit of platelet concentrate. Postoperatively she was having continuous oozing from the abdominal wound. There was no excessive vaginal bleeding. In spite of giving single donor platelets and 21 units of platelet concentrate there was no improvement in her condition.

On the fifth postoperative day 2.4 mg (40 microgram/kg) of rFVIa was given intravenously and a dose of 1.2 mg was repeated after 24 hours. On the sixth postoperative day the oozing from the wound stopped. Subsequently she had an uneventful postoperative period and the baby and mother were discharged on the 10th postoperative day. At 6 weeks postpartum, the baby had normal bleeding time, clotting time and clot retraction time with platelets count 1.8 lakh. Mother however had bleeding time of 10 minutes with normal platelet counts.

Discussion

Glanzmann's thrombasthenia was first described in 1918. There are different varieties of the disease which is associated with postpartum hemorrhage[1]. Essential diagnostic features are

1. Normal platelet count and morphology with prolonged bleeding time
2. Absence of platelet aggregation in response to ADP, collagen, epinephrine, thrombin and normal ristocetin induced aggregation.
3. Diagnosis is confirmed by measuring the amounts of platelet glycolproteins GpIIb/IIIa on the platelet surface membrane using monoclonal antibodies.
4. Coagulation studies such as prothrombin time and activated partial thromboplastin time are normal.

Bleeding in thrombasthenia requires transfusion of normal platelets. Because bleeding is a lifelong problem use of HLA matched platelets should be considered to lessen the chance of platelet alloimmunization, which can limit the effectiveness of transfused platelets.

Recombinant factor VII has been shown to be effective in treatment of severe bleeding episodes and for coverage of surgical procedures in patients with previous history of ineffective platelet transfusion. Factor VIIa is given to activate the extrinsic pathway of the coagulation cascade[1]. Meanwhile the management of the disease consists of avoidance of trauma and medicines such as aspirin, which may further affect platelet function and treatment of major bleeding episodes by platelet transfusion.

Heavy periods may respond to hormone therapy. Our patient continued to have unabated oozing from the abdominal wound in spite of receiving multiple platelet transfusions. It is possible that the platelets may have become ineffective due to alloimmunization. This was evidenced by the sudden improvement in her condition when rFVIIa was given. This case is reported because it is a rare disorder and it is even more rare to encounter it in pregnancy. Excessive bleeding in such patients is treated with large quantities of platelet transfusion. However this case highlights the importance of the treatment with rFVIIa in patients who do not respond to platelet transfusion.

References

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