Glanzmann's Thromboasthenia in Pregnancy

SP Aher, Ashok Kumar, D Pardeshi, G Gawli, N Rao, K Rawat

Abstract

Glanzmann's Thromboasthenia is a congenital acquired platelet disorder with an autosomal recessive mode of inheritance. It is a haemorrhagic disorder characterized by severe reduction or absence of platelet aggregation in response to multiple physiological events due to qualitative and quantitative abnormalities of platelets glycoproteins GP IIb and GP IIIa. Pregnancy and delivery are rare and have been associated with a high risk of severe postpartum haemorrhage.

Introduction

Glanzmann's thromboasthenia is a inherited disorder of platelet function characterized by bleeding episodes. The laboratory studies showed prolonged bleeding time with absent or decreased clot retraction and normal platelet counts. Coagulation studies are normal. Platelet aggregation in response to agonist. ADP, collagen, arachidonic acid is absent. Clinically patient present with symptoms like purpurae, epistaxis, gingival haemorrhage and menorrhagoea, post partum haemorrhage.

Case Report

A 22 year female patient diagnosed case of Glanzmann's thromboasthenia with 1st trimester of pregnancy was referred for further management. Past history of menorrhagia, bleeding gums since 10 years. History of chronic anaemia since 10 years for which patient was on oral iron supplements and her heavy prolonged menses was controlled with tranexamic acid. History of epistaxis on and off in past which was controlled with nasal packing. History of easy bruising, and ecchymosis all over the body. No other history of major illness like Kochs/ hypertension/DM in past or in family members. History of one first trimester abortion which was spontaneous with no complications - 1 year back.

Investigations

Hb = 6.2 gm%

WBC = 3,800

Haematocrit of 25%

Pl = 2,96,000

Bleeding time was prolonged = more than 15 minutes

No agglutination noted with adenosine-di-phosphate, adrenaline, collagen and arachidonic acid in presence of ristocetin.

Renal/liver function test were normal.

Platelet fragility tests

Count = 196 µg/L

MPV = 10.50 fl

BT = Duke method = 3.05 min

Clot retraction = 27% at 1 hr

Pl. aggregation ADP = 2.5 µm/L, APD = 5%

Epinephrine, 2 µm/l = 1%

USG = Single viable foetus

Collagen = 2%

Ristocetin (1.5 mg/ml) = 75%

Peripheral smear = Pl aggregation not seen except of ristocetin.

Patient diagnosed as glanzmann's thromboasthenia with pregnancy.

Course and treatment given

Patient was closely monitored by Haematology and obstetric units. Patient was monitored every monthly for platelet counts, anti-platelets antibodies, intrauterine intracranial haemorrhage; anti HLA and GP IIb/IIIa or GB Ib iso antibodies. Patient had uneventful full term normal vaginal delivery. Her platelet counts were kept to near target levels with DDAVP (desmopressin) and antifibrinolytic agents.

Department of Haematology, Nair Hospital, Bombay Central, Mumbai - 400 008.

Tranexamic acid was given at dose of 10 mg/kg tds and DDAVP desmopressin was given as IV infusion - 0.2 µg/kg diluted on 50 ml of normal saline over 30 min at the time of delivery. Thus the risk of maternal haemorrhage was avoided and rFVIIa was kept as standby treatment in case of emergency. Mother and child were discharged after 3 days on oral tranexamic acid (10 µg/kg) for 15 days.

Discussion

Pathophysiology : Receptors GP IIb and GP IIIa required for platelets aggregations are absent or reduced resulting in fracture of platelet plug formations or cloth retraction at vascular site.

1. Antifibrinolytic agents e.g. tranexamic acid is given orally at the dose of 15-25 mg/kg. This is to reduce active bleeding episode. It can be given as mouthwash (10 ml of 5% solution qds).
2. Desmopressin (DDAVP) - this drug is used to increase the levels of circulating VWF. It is used when tranexamic acid alone is ineffective. It causes fluid retention, hyponatraemia. Fits are to be avoided in patient < 2 years of age and in atherosclerosis. It is given as - IV infusion - 0.2 µg/kg - 0.3 µg/kg infusion diluted in 50 ml of normal saline over ½ hr.

   Subcutaneous infection - is also given at rate of 0.3 µg/kg.
3. Intranasal spray - 150 µg per dose (0.2 - 0.3 µg/kg) used in active bleeding disorder.
4. Platelet transfusion - are used when the above treatment fails. Patient with repeated platelet transfusions develop alloantibodies either to HLA antigens or missing GPs.

   i) Random donor platelets - one adult dose is equivalent to 4-6 single donor units given in dose of 10-15 ml/kg.

   ii) HLA selected platelet transfusion.
5. Plasma exchange can be used to reduce antibody titres.
6. Recombinant factor VII a (rFVIIa) is an alternative therapeutic agent.

Pregnancy with Glanzmann's thromboasthenia should be monitored antenatally for development of anti HLA and GP IIb/IIIa or GP Ib iso antibodies. The normal foetus is at risk of alloimmune thrombocytopenia as result of transplacental haemorrhage followed by sensitization. Antiplatelets antibodies cross the placenta and cause neonatal thrombocytopenia, intrauterine foetal haemorrhage. Neonates are to managed with platelets transfusion, ABO and Rhesus identical, CMV screened HLA matched platelets.

Haemopoietic stem cell transplantation is the last appropriate form of therapy with stem cells transplantation.

INHIBITING INTERLEUKINS TO REDUCE ASTHMA SYMPTOMS

Evidence that T helper (Th) 2 cytokines such as intereukin 4 or interleukin 13 have a crucial role in the onset and development of clinical asthma is scarce. Pitrakinra was administered via two routes - by subcutaneous injection once daily or by nebulisation twice daily. The investigators showed that decreases in FEV1 after allergen challenge were significantly attenuated after 4 weeks of inhalation of pitrakinra.

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