

CASE REPORT

PREGNANCY WITH PLATELET FUNCTION DISORDER

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ABSTRACT: Platelets play a vital role in haemostasis. Antenatal patients with platelet function disorders should be managed in tertiary care centres that are well equipped to tackle any obstetric haemorrhage that can ensue during labour and delivery. Primi gravida was admitted for safe confinement. She had been evaluated earlier for complaints of multiple episodes of mucosal bleeding. On evaluation she had normal platelet counts and coagulation factor assay was normal. Platelet aggregometry revealed mild disorder of platelet aggregation. She was planned for induction of labour after arranging enough blood and blood products. She got into active labour and was put on syntocinon augmentation. She had emergency Caesarean section for foetal distress. Oxytocics were given proactively. Intraoperatively platelet transfusions and tranexamic acid infusion were given. Complete haemostasis was achieved. She had an uneventful postoperative period. Patients with functional platelet disorders can be successfully managed with local application of antifibrinolytic agents like tranexamic acid, in case of minor bleeds. Platelet transfusions are very effective in tackling major bleeds, especially during surgeries and for obstetric indications. If a patient has the history of clinically significant bleeding suggestive of platelet dysfunction, appropriate platelet function tests should be obtained so that the risk of bleeding can be adequately assessed and therapy chosen more rationally. . In obstetric practice the response of such patients to platelet transfusions has been excellent.

INTRODUCTION: Platelets play a key role in both hemostasis and thrombosis. The accurate assessment of platelet function is vital for identifying patients with platelet dysfunction or hyperfunction. The ability to test platelet function in the routine laboratory has been revolutionized with the introduction of platelet aggregometry. The availability of newer and more reliable platelet function analyzers, flow cytometry and molecular biological techniques is changing our approach to platelet function analysis. Antenatal patients with platelet function disorders should be managed in tertiary care centers that are well equipped to tackle any obstetric hemorrhage that can ensue during labor and delivery.

CASE HISTORY: Mrs. X a 29 year old primi gravida at 39 weeks 5 days gestation was admitted for safe confinement. She had conceived spontaneously. Dating scan was done which confirmed the dates. NT scan was done and first trimester screening was negative. Anomaly scan was done which ruled out anomalies. Blood parameters were done serially and were found to be normal. Growth profile scan was normal. She did not have history of any bleeding manifestations throughout the pregnancy. Antenatal period was uneventful.

PAST HISTORY: She had history of bleeding gums from childhood. She had history of spontaneous episodes of ecchymosis since the age of 8 years. She had history of hematuria for about 5 years from

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the age of 6 years and again recurring in between with the last episode occurring one year back. She had history of recurrent episodes of spontaneous bilateral epistaxis from the age of 18 years. She had regular menstrual cycles with no history of menorrhagia in the past. She did not require any blood transfusions and had not been on any specific medications in the past. There was no history of any bleeding diathesis in her family. She was evaluated at the age of 21 years for any possible bleeding disorder. On evaluation she was found to have normal baseline blood parameters. (Hb-11.1g%, BT 3 minutes, PT, PTT and INR were normal. Thrombin time was normal with good clot retraction. Serum fibrinogen level was normal. Platelet count was normal and platelet morphology was normal). The activity of clotting factors (Factor VIII, Factor IX, Factor XI, Factor XIII, vWF: RCo) were found to be normal. Platelet aggregometry showed normal response to normal dose (1.5mg) and low dose (0.5mg) of ristocetin. Normal responses were noted to collagen and arachidonic acid also. ADP (5micromoles/litre) and (20micromoles/litre) showed only primary wave of aggregation followed by disaggregation. Subnormal response was noted to epinephrine (10micromoles/litre).

Lumiaggregometry using collagen and thrombin as agonists showed a normal response. Absent responses were noted to TRA (thromboxane receptor analogue) and TRAP (thromboxane receptor activating peptide). Her hepatic and renal function tests were normal. ANA panel was negative. Total complement level, C3 and C4 levels were normal. The complete coagulation workup revealed mild platelet dysfunction. She was advised topical application of tranexamic acid paste in case of minor superficial bleeds and random donor platelet transfusions in case of major bleeds, surgeries and obstetric indications to achieve hemostasis.

MANAGEMENT OF LABOUR AND DELIVERY: She was planned for induction of labor at 40 weeks with PGE2 gel after reserving 2 units of packed cells, 2 units of FFP and 6 units of platelets. She got into labor spontaneously. ARM was done followed by syntocinon augmentation. 3 units of platelets were transfused in labor. She was monitored adequately and was put on continuous fetal heart rate monitoring. She was taken up for emergency LSCS 5 hours after starting syntocinon augmentation in view of fetal distress (persistent fetal heart rate decelerations upto 90 bpm). Baby was fine (boy, 3.36 kg with good Apgar score and cord pH of 7.360). Intraoperatively she was started on oxytocics (syntocinon IV and IM along with IV methergine, IM PGF2 alpha and PGE1 per rectally) to enhance uterine contractility. She was given 6 units of platelets and 1 unit of packed red cells intraoperatively. She was given tranexamic acid 1 gram IV infusion intraoperatively. Uterus was closed in two layers after ensuring perfect hemostasis. There was no atonicity of the uterus or excessive bleeding from the surgical site. Intraperitoneal drain was kept and abdomen was closed in layers. Postoperatively she was continued on tranexamic acid infusion 1gram intravenously TDS along with syntocinon infusion, parenteral antibiotics and analgesics. She was observed and monitored carefully in the high dependency unit for 24 hours. She was given 3 units of platelets each on postoperative days 1 and 2. The drain was removed on the second day and she was discharged on the third day. On follow up she was found to have adequate wound healing with no morbidity related to the platelet dysfunction.

DISCUSSION: Effective hemostasis requires the orderly interaction of components of the blood vessel wall with platelets and circulating plasma proteins. Hemostatic defects lead to hemorrhage by interrupting these interdependent reactions. The initiating event is an injury that damages or removes the vascular endothelial lining and exposes the subendothelial connective tissue.¹The

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optimal adherence of platelets to the exposed collagen fibres requires a specific plasmaprotein called Von Willebrand's factor that adheres to both.²The adherent platelets then degranulate, releasing ADP which recruits additional platelets to aggregate upon the adherent platelet monolayer.² Aggregation results from the binding of fibrinogen to receptor sites on the platelet glycoprotein(Gp 2b-3a complex).This process rapidly produces the platelet plug. After activation, platelets release potent vasoconstrictors like thromboxane A₂ which induce further platelet aggregation. This is followed by plasma coagulation.

Patients are often encountered with normal platelet counts but still exhibiting clinical manifestations suggestive of a defect in primary hemostasis. In such patients, the hemostatic abnormality can arise from a defect in platelet adhesion or aggregation or even a defect in release reaction. Estimation of bleeding time as well as platelet aggregation studies are useful. Platelet aggregation study is an in vitro test in which platelet plug formation is simulated by incubating stirred platelet suspension with chemicals known to cause platelet aggregation and release like ADP, collagen, epinephrine, ristocetin and arachidonic acid.

Occasionally patients have a defect in primary platelet aggregation called Glanzmann's thrombasthenia where platelets are totally unable to aggregate to any stimulus. This is extremely rare and is due to selective loss in platelet fibrinogen receptors located on Gp 2b-3a complex. Failure to complete the aggregation sequence by undergoing release and secondary aggregation is a much more common defect. Such failure results from a deficiency of arachidonate metabolism or from a deficiency in stored ADP in the platelets. Patients with either defects have prolonged bleeding time and platelets that do not aggregate normally when challenged with agents such as epinephrine or collagen, but will respond to larger doses of ADP (10micromoles/litre).Congenital or acquired defects of platelet function normally affect aggregation with ADP, epinephrine, collagen and arachidonic acid and do not affect aggregation with ristocetin whereas in von Willebrand's disease, ristocetin induced aggregation is reduced.³⁻⁵

Patients with functional platelet disorders do not usually have serious bleeding episodes. They may bleed only when seriously challenged with trauma or surgery. Most of them do not require specific therapy. They usually respond to platelet transfusions in obstetric indications with improved clinical hemostasis. In cases of mild superficial bleeding or following minor surgical procedures, antifibrinolytic agents like tranexamic acid has been found to be useful in them to reduce bleeding⁶

SUMMARY: If a patient has the history of clinically significant bleeding suggestive of platelet dysfunction, appropriate platelet function tests should be obtained so that the risk of bleeding can be adequately assessed and therapy can be chosen more rationally. Therapeutic decisions should not be based on the tests alone as they may not predict clinical bleeding always. In obstetric practice the response of such patients to platelet transfusions has been excellent.

REFERENCES:

1. Peter W Marks, Robert I Hande. Bleeding and thrombotic disorders. Office Practice of Medicine 4th edition 2008; 59:756.
2. George JN, Shattil SJ. The clinical importance of acquired abnormalities of platelet function. N Engl J Med 1991; 27:324.

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3. Hayward CP, Pai M, Liu Y, et al. Diagnostic utility of light transmission platelet aggregometry: results from a prospective study of individuals referred for bleeding disorder assessments. *J Thromb Haemost* 2009; 7:676.
4. Cattaneo M, Hayward CP, Moffat KA, et al. Results of a worldwide survey on the assessment of platelet function by light transmission aggregometry: a report from the platelet physiology subcommittee of the SSC of the ISTH. *J Thromb Haemost* 2009; 7:1029.
5. Quiroga T, Goycoolea M, Matus V et al. Diagnosis of mild platelet function disorders. Reliability and usefulness of light transmission platelet aggregation and serotonin secretion assays. *Br J Haematol* 2009; 147:729.
6. Bolton- Maggs PH, Chalmers EA, Collins PW, et al. A review of inherited platelet disorders with guidelines for their management on behalf of the UKHCDO. *Br J Haematol* 2006; 135:603.

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