



BERNARD-SOULIER SYNDROME IN PREGNANCY: CASE SERIES

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ABSTRACT

Bernard-Soulier Syndrome is a rare syndrome, first discovered in the year 1948. The pattern of inheritance is autosomal recessive. It primarily causes platelet dysfunction. Rarely, an autosomal dominant form has also been described. We present a series of two cases, both of whom delivered with us. Management was medical, with transfusions of blood products and platelets.

KEYWORDS : Bernard- Soulier, Pregnancy, syndromes in pregnancy, platelet counts, blood transfusion in pregnancy

INTRODUCTION

Bernard-Soulier syndrome is clinically characterized by a decrease in platelet counts, large giant platelets and a prolonged bleeding time. The primary dysfunction is due to genetic defects in the formation of the glycoprotein complex [Ib/IX/V]. This is the von Willebrand factor receptor on the platelet surface. Von Willebrand factor acts as a bridge between the sub-endothelial matrix and the platelets.

The presentation can be varied ranging from no symptoms to mild bleeding episodes like epistaxis, menorrhagia, easy bruising to bleeding even with little trauma (1). Bernard-Soulier complicating pregnancy is very rare. Most common cause for thrombocytopenia in pregnancy is gestational thrombocytopenia. This case series aims to raise awareness about the rare cause for thrombocytopenia in pregnancy i.e., Bernard-Soulier Syndrome which is a clinical challenge for both Haematologists and Obstetricians (2).

CASE REPORT

Mrs. K presented to our antenatal clinic as a unbooked Primi at 38 weeks of gestation with complaints of abdominal pain and show. All of her antenatal period was uneventful. Her menstrual history was normal and there was no past history of any bleeding episodes. She was diagnosed with Platelet dysfunction at a tertiary institution outside, the details of which were unavailable. She was a known case of chronic migraine on treatment with Flunarazine.

On examination, she was conscious, oriented and afebrile. Her vitals were stable. Cardiovascular system and respiratory system were normal. On abdominal examination, the uterus corresponded to term size, mild contractions were present and the presentation was cephalic. Fetal heart rate was good. Per vaginal examination revealed a soft cervix, which was posterior and 50% effaced. Her os was dilated to 1 cm.

As her platelet count was low (75,000) with giant platelets in smear study, after consultation with a Haematologist, three single donor platelets were transfused. Repeat platelet count was 1.2 lakhs. She was induced with one Dinoprostone gel and later amniotomy was done. Oxytocin augmentation of labour was started. As the CTG trace was abnormal, patient was taken up for emergency LSCS. Seven Platelets and one packed cell were transfused intraoperatively. There was increased bleeding from the incision site. There were no postoperative complications. Postoperatively the patient was transfused three Platelets. Drain was removed on 3rd postoperative day. Wound was healthy. Baby with mother on direct breast feeding hence patient was discharged on fifth postoperative day. Patient was doing well during follow-up.

CASE REPORT 2

Mrs.S, a booked Primi at 38 weeks+ 3 days with Bernard-Soulier Syndrome complicating pregnancy was admitted for safe confinement. No abdominal pain/ leaking/ bleeding per vaginum. Perceives fetal movements well.

Her first and second trimesters were uneventful. There was a history of increased BP from 33 weeks for which patient was started on Alpha

methyl dopa. She was diagnosed with Bernard-Soulier Syndrome in her childhood after repeated episodes of bleeding gums. She has a history of four blood transfusions in the past. Her menstrual history was normal. One of her brothers suffers from Bernard-Soulier Syndrome.

On examination, patient was conscious, oriented and afebrile. Her vitals were stable. Cardiovascular and respiratory system were normal. On abdominal examination, uterus was term size, relaxed and the fetus in cephalic presentation. The fetal heart rate was good and liquor was adequate.

All baseline investigations were done which was found to be normal. Platelet counts were normal but smear study showed giant platelets and increased bleeding time. Previously done platelet aggregometry revealed findings consistent with BSS. As per hematologist opinion, patient was transfused with 2 units of single donor platelets and then induced with 2 doses of Dinoprostone gel. Patient was transfused with another two units of single donor platelets while in labour to prevent postpartum hemorrhage. Amniotomy revealed grade 2 meconium stained liquor but the CTG trace remained normal. Patient delivered by normal vaginal delivery with episiotomy. The 1 and 5 minute APGAR were 8 and 9.

Postnatally patient was transfused with two more units of single donor platelets. Patient was discharged on postnatal day three at request. during follow up, she was doing well.

DISCUSSION

Presentation of Bernard-Soulier with pregnancy is rare hence there are no management protocols evolved. During review of literature, all but one patient was found to have severe postpartum hemorrhage. It was also of the secondary type. Antepartum Hemorrhage is rare and self-limiting(3). Platelet transfusions form the mainstay of the treatment, both prophylactic and therapeutic. Other modes include whole blood transfusion, using antifibrinolytics.

Intravenous immunoglobulin and steroid therapy have also been described but were not used in these patients due to less severity of symptoms and available of single donor platelets. Some patients might require intranasal desmopressin (DDAVP), recombinant factor VIIa due to formation of antibodies in the recipient's blood to the platelet receptors, following repeated platelet transfusions. Hence, when patient does not respond to platelet transfusions, the above modalities may be used (4,5).

There are no recommendations for the mode of delivery. Caesarean sections are reserved for obstetric indications(3,4). Obstetric hysterectomy following postpartum hemorrhage is rare but has been reported(6). These patients bleed typically, following development of alloantibodies.

Neonates are usually unaffected but may develop bleeding secondary to alloimmune thrombocytopenia. The mother produces antibodies to the fetal platelets with intact functioning glycoprotein receptors(7,8).

Pregnancy complicated by this syndrome needs to be managed by a multidisciplinary team- Hematologist, Obstetrician and Fetal Medicine Specialists. Other complications should be concurrently treated, like anaemia. Platelet transfusions should be HLA matched. Regional Anesthesia is contraindicated. Genetic counseling should be offered.

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