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Successful Outcome of Pregnancy in Bernard - Soulier Syndrome with Temporary Endovascular Balloon Occlusion of Uterine Arteries: A Case Report

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Abstract

An uncommon congenital bleeding illness known as Bernard-Soulier Syndrome (BSS) is primarily inherited in an autosomal recessive pattern. Depending on the individual mutation, the phenotype of BSS varies significantly. There is no consensus on the best treatment for patients with BSS, and pregnancy is linked to a high risk of bleeding for both the mother and the newborn. The use of perioperative endovascular balloon occlusion of the internal iliac arteries (uterine artery) is a minimally invasive technique that reduces blood loss and the need for transfusions in high bleeding risk patients, such as those with BSS.

This case describes a successful pregnancy outcome with endovascular intervention in a woman with BSS who was closely monitored during pregnancy, the peripartum period, and the postpartum period, and who had a planned birth. Minimally invasive and effective endovascular prophylactic measures against bleeding reduce blood loss, the need for transfusions, platelet refractoriness, and hospital stay during a cesarean section in patients with BSS.

Keywords: Bernard-Soulier Syndrome, Blood Transfusion, Blood Platelet Disorders, Pregnancy.

Introduction

Bernard-Soulier Syndrome (BSS) was initially identified in 1948 in a young patient who exhibited hemorrhagic symptoms, prolonged bleeding time, thrombocytopenia, and unusually large platelets.[1] This syndrome is inherited in an autosomal recessive manner and results from abnormalities within the platelet membrane glycoprotein (GP) Ib-IX-V complex. These abnormalities impair platelet adhesion and prevent platelets from binding effectively to Von Willebrand factor and thrombin.[3] Common clinical manifestations include gingival and cutaneous bleedings, recurrent episodes of epistaxis, menorrhagia, peri & postoperative bleeding, and bleedings related to trauma. The severity of bleeding varies depending on the underlying mutation.[2]

Neonatal alloimmune thrombocytopenia, blood transfusion, hysterectomy, and primary and secondary postpartum haemorrhage are all risks that are increased by BSS pregnancy. There is controversy on the appropriate way to handle these instances because there is not enough of data in the literature. In this case report, a Pakistani tertiary care hospital's management of BSS during pregnancy is discussed.

Case Report

A 28-year old Asian primigravida from Sindh Pakistan with BSS presented to her obstetric appointment at 36+5 weeks of gestation. She remained asymptomatic in pregnancy. The diagnosis of BSS was made at the age of 20 years on CBC, peripheral film and platelet aggregation studies. The patient had history of excessive bleeding after trauma, heavy menstrual bleeding, and platelets transfusion once during her lifetime. She had no other past medical history and had not undergone any surgery. There was no history of tobacco, alcohol or other substance abuse. Her physical examination was unremarkable, and gynecological exam showed fundal height of 36 cm and fetal heart rate of 145 bpm. She had severe thrombocytopenia (platelet count 16 $x10^9$/L), with presence of giant platelets in peripheral blood smear. Her preoperative hemoglobin was maintained above 10 g/dl. Other laboratory tests were unremarkable for any pathology.

Multidisciplinary discussion including gynecologist, hematologist, interventional radiologist and anesthetist was conducted. Elective cesarean section under general anesthesia, with ballooning of uterine artery and transfusion of single donor platelets during surgery

were planned. Written consent for c-section delivery was obtained from patient. Platelets, red blood cells, and activated factor VII were arranged as a backup for the patient in case of uncontrolled bleeding. Tranexamic acid 1 g thrice a day was prescribed for use one day prior and two days after the procedure.

The patient was admitted at 37+5 weeks of gestation. On the following day, at 37+6 weeks, an interventional radiologist performed the inflation of both uterine arteries before the scheduled cesarean section. The procedure began with standard disinfection, followed by the administration of local anesthesia using 2% lidocaine. Once the right and left femoral arteries were successfully punctured using the Seldinger technique, 7Fr arterial catheter sheaths were inserted on both sides. Subsequently, a balloon catheter was carefully positioned in the anterior division of the internal iliac arteries on both sides, guided by a wire. Contrast was injected in small amount to confirm the position of catheters followed by arterial sheaths fixation. She underwent cesarean section with pffenesteil incision under general anesthesia. Just before giving the surgical incision, the balloons were inflated in anterior divisions of internal iliac arteries bilaterally, sizes of balloons were 6*60 mm on left side and 5*60 mm on right side with atm pressure 10 on both sides. Single donor platelets were transfused intraoperatively. An alive and healthy female baby, cephalic, 3 kg with APGAR scores 8/1 and 9/5 was born. Prophylactic uterine and vaginal packing done. She remained stable hemodynamically throughout the procedure with minimal blood loss (EBL 500 ml).

Balloons were deflated after 3 hours of procedure. Femoral sheaths, uterine and vaginal packing's were removed after 24 hours of procedure. In total, patient received single donor platelets on the day of procedure and 6 random donor platelets on 1st post operative day. Tranexamic acid was given for 24 hours after procedure. She did not experience primary and secondary postpartum hemorrhage and surgical wound healed without any complication. The patient was discharged on 4th post operative day. Baby was fine and there was no episode of bleeding in neonatal period and she was discharged with her mother.

Discussion

Bernard-Soulier Syndrome is a rare platelet disorder inherited most often as recessive trait. An autosomal dominant trait is also described.[1] Due to diverse phenotypic presentation and nonspecific symptoms of BSS, age at diagnosis differs widely from patient to patient. Affected females are usually diagnosed after menarche, with a mean age of 19 years in a recent review.[2] Our patient presented with history of excessive bleeding after trauma and episodes of heavy menstrual bleeding at the time of diagnosis.

In BSS, pregnancy not only has a diverse course for various patients, but also a variable outcome for the same patient between pregnancies.[2,4] These women may require an emergency hysterectomy due to their elevated risk of intrapartum and postpartum haemorrhage. Maternal antiplatelet antibodies have been linked to an increased risk of alloimmune neonatal thrombocytopenia, which can result in cerebral haemorrhage in the newborn.[6] Due to these reasons delivery at tertiary care hospital is advised. Mode of delivery in these patients is controversial, with no significant difference in outcomes of pregnancy between women delivering vaginally or by cesearian section. Mostly the mode of delivery is decided by the obstetrician considering the history of patient, consulting multidisciplinary team including hematologist and anesthetist as well as patient's preference. Our patient did not opt for the trial of normal vaginal delivery. General anesthesia is the choice for these patients due to risk of epidural hematoma with regional anesthesia.[2,10] Peripartum period is the most important time of pregnancy in such women, so mode of delivery should be planned and prophylactic measures against bleeding should be taken in advance. Tranexamic acid, desmopressin, recombinant factor VIIa, platelet transfusion, and uterine artery blockage can all be used to prevent bleeding. As this is the most effective and secure precaution before surgery for individuals with congenital platelet abnormalities, our patient underwent a single-donor platelet transfusion.[9,10] Nevertheless, patients who get platelet transfusions run the danger of becoming alloimmunized, which can make subsequent platelet transfusions ineffective.[6,8]

We opted for balloon occlusion of the uterine arteries to prevent excessive bleeding during surgery and to mitigate the risk of postpartum hemorrhage. This strategy proved successful, as there was no excessive bleeding during the surgery, and the patient required only a single donor platelet transfusion intraoperatively. There was no need for a blood transfusion during or

after the surgery. Close vigilance is required for at least six weeks postpartum due to the risk of secondary PPH.[7,8] However, our patient's postpartum period was uneventful.

Conclusion

BSS is a rare bleeding disorder that can complicate pregnancy, and its course is variable and unpredictable. A minimally invasive and effective endovascular prophylactic measure against bleeding reduces blood loss, transfusion requirements, platelet refractoriness, and hospital stay during cesarean sections in these patients

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