Bernard-Soulier syndrome: A challenge for anesthetist in an emergency surgery

Sir,

Bernard-Soulier syndrome (BSS), reported in 1948 for the first time,[1] is a rare hereditary cause of bleeding diathesis characterized by platelet dysfunction secondary to the absence of the von Willebrand factor receptor on the platelet surface.[1]

We present the case of a 20-year-old woman who was diagnosed with BSS at the age of 4 years. Her history revealed on and off episodes of epistaxis, gum bleeding and poly-menorrhagia mandating platelets transfusion on three different occasions during her life time. Beside this conservative management, she did not receive any further treatment for this syndrome.

At 38 weeks of gestation, due to her suboptimal continuous tocograph, an emergency cesarean section was planned. Her preoperative anesthesia assessment revealed a smooth antenatal course without any major bleeding episode. The laboratory investigations revealed hemoglobin of 10.2 g/dl with hematocrit of 30.4, white blood cell count of 12,000/cu mm and unrecordable platelet count. Request was made to the laboratory for manual counting of platelets, which then showed the figure of 155,000/cu ml of blood. This discrepancy in two different methods of platelet counting is quite typical of BSS because of the increase size of platelets (giant platelet syndrome).[2] The coagulation profile showed prothrombin time 9.3/11.0, activated partial thromboplastin time 25.2/30.0, and international normalized ratio 0.88. Other laboratory findings were within normal ranges including bleeding time (2 min) and clotting time (4 min). Based on these findings, general anesthesia was planned after detailed informed consent.

As per advice of a hematologist, 10 units of platelets, two units of packed red blood cells and arrangement of factor VII was made for handling crisis situation. Since major intraoperative and postoperative hemorrhage was the primary concern, two large bore intravenous cannula of 14 G was inserted, and six units of platelet and 2 g of tranexamic acid was given intravenously before taking the patient in operating room.

In the operating room, baseline monitoring was applied which included pulse oximeter and electrocardiogram. An arterial line was secured in left radial artery for measuring blood pressure. Preoxygenation for 3 min with 100% FiO_2 was followed by rapid sequence induction utilizing sodium thiopental (5 mg/kg) and succinyl choline (1.5 mg/kg). Trachea was intubated in first attempt and anesthesia was maintained with isoflurane 1% in oxygen (40%) and nitrous oxide (60%). Paralysis was maintained with atracurium to aid surgery. A healthy baby girl was delivered with good APGAR scores of 8/1 and 9/5 respectively. Morphine 7 mg was used once baby was delivered, and it was supplemented with 1 g intravenous paracetamol.

The patient remained hemodynamically stable throughout the procedure. There was total blood loss of 900 ml. Patient was successfully extubated in the end.

Postoperatively in postoperative anesthesia care unit, she was transfused with six platelets units and tranexamic acid was started at a dose of 500 mg every 6 hourly intravenously for further 48 h, that were later switched to oral regime. Platelet count postoperatively was 76,000/cu ml of blood. Patient was discharged home uneventfully on 4th postoperative day.

Till date, only some case reports about the pregnant patients with BSS have been published, among them the detailed reports about anesthetic management of such patients are very few. In this report, the authors aim to emphasize on the clinical course, the anesthetic management and the need for multidisciplinary team approach at tertiary care centers in such cases for successful outcomes. Pregnant women with BSS should ideally be managed in a tertiary unit with thorough hemostatic assessment of the patient. The clinical expertise and readily available access to platelet and other blood products should be ensured in all cases during delivery or cesarean section to minimize the chance of life-threatening hemorrhage.

Platelet transfusion remains the mainstay of treatment and has been used both prophylactically and therapeutically.[3] Antifibrinolytic drugs such as aminocaproic acid or tranexamic acid use have a controversial role in literature. The other treatment modalities are

1-deamino-8-d-arginine vasopressin, recombinant factor VIIa, steroids and intravenous gamma globulin plasmapheresis.[4]

Report of further cases and individual experience in managing these pregnancies will be helpful to obtain better knowledge. An international registry for BSS in pregnancy is desirable in this regard.

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Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/0970-9185.161709