

Pregnancy with Bernard Soulier Syndrome – A Case Report

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Bernard Soulier syndrome is a very rare congenital recessive disorder characterized by severe bleeding tendency. There is reduced platelet adhesion owing to deficiency of platelet glycoprotein IIb/IIIa receptor. The only available treatment of these patients is either the use of platelet transfusion in the case of bleeding episode or as prophylaxis prior to surgery.

A 34-year-old female patient presented to our clinic on 12/6/99 at 36 weeks gestation with severe pruritus as a known case of Bernard Soulier syndrome. She was advised for placental transfusion and safe confinement. This patient had history of menorrhagia, gum bleeding, epistaxis and ecchymosis bleeding from cuts since early childhood. Her only sister and one cousin brother had similar illness. There was history of consanguineous marriage in the family. She had been diagnosed in ALMG in 1996. She had received blood transfusions in the past and was advised against pregnancy. She discontinued the previous treatment for primary infertility and came to the O.R. in the last trimester.

Examination showed a 36-year-old female patient, normal appearing woman with gum hyperplasia. CBC & PT, and renal/liver systems were within normal limits.

Investigation: Hb 7.4 g/dl, Platelet count 10000, platelet giant platelets seen, APTT 49 sec, Bleeding time 7 minutes, Clotting time 3 minutes 30 seconds, Prothrombin time 11 seconds, DVT-negative, HLA-Ag Negative, VDR non-reactive.

Patient went into labour on the day of admission and had emergency C-section. A 3.6 kg male child of 38 weeks was born. The patient was managed with fresh blood transfusion (total 4 units), Aspirin 80 mg, Streptokinase injection and antibiotics. She also received Wycorone tablets (Warfarin) daily 5 mg along with Tab. heparin 10 mg twice daily. The Wycorone was gradually tapered off over next 2 weeks. The 7th post-operative platelet count was 45,000/mm³ and Hb 10 gm/dl. She was discharged on 1st post-operative day, remaining at the ward did not show any bleeding tendency so far.

Bernard Soulier syndrome is a rare congenital condition and its association with successful pregnancy outcome is very rare.