

Case report

Successful maternal and fetal outcome of a patient with paroxysmal nocturnal hemoglobinuria in pregnancy: a case report

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Abstract

Paroxysmal nocturnal hemoglobinuria (PNH) is an acquired hemolytic anemia characterized by paroxysmal intravascular hemolytic episodes, hemoglobinuria, thrombocytopenia and thrombotic tendency especially during stressful conditions like pregnancy. We report the case of a second gravida known to have PNH on prednisolone therapy. She was booked in early pregnancy and had regular ANC follow-up. She required repeated blood transfusions throughout pregnancy for severe anemia and thrombocytopenia. She had a spontaneous preterm delivery at 32 weeks and delivered a live baby weighing 1.4 Kg. Both the mother and neonate were discharged in good condition.

Keywords: Paroxysmal nocturnal hemoglobinuria, pregnancy, steroid therapy.

Introduction

Paroxysmal nocturnal hemoglobinuria (PNH) is an acquired hemolytic anemia caused due to a defect of glycosylphosphatidylinositol (GPI) anchored proteins in the cell membrane of bone marrow stem cells. This causes increased sensitivity of the red cells to complement which in turn leads to intravascular hemolysis. Periods of hemolysis can be evoked by various agents namely infections, transfusion, iron intake, menstruation, surgery and even pregnancy. PNH is characterized by hemoglobinuria, anemia,

thrombocytopenia, and thrombosis especially of abdominal vein^[1] and an increased risk of infections in addition to paroxysmal hemolytic episodes. PNH affects mainly adults. The disease has a chronic remitting course with a median survival of 10-15 years.^[2] Maternal mortality estimates range from 5.8% to 20.8%. Most common cause of death is thromboembolism.^[3, 4] These patients are relatively subfertile and many pregnancies end up as abortions or premature deliveries. There are few case reports of patients with PNH having a good pregnancy outcome. We present the case of a second gravida with PNH, treated with several blood transfusions and steroids during pregnancy who had a successful pregnancy outcome

Case Report

The patient was a 26 year old second gravida who was known to have PNH since her last pregnancy 2 years back. During her first pregnancy the patient developed severe anemia and started passing black urine. Hemoglobin was 6.7 g%, platelet count 70,000/dl and total and differential leukocyte counts were normal. No specific diagnosis was made. She received several units of packed cell transfusions but the hematological abnormalities persisted. She delivered 4 weeks later at 29 weeks. However, the baby succumbed due to prematurity. She received transfusion of red cells and platelets during delivery. Hemoglobin and platelet counts failed to rise in the postpartum period. She was then subjected to investigations and the diagnosis

was verified by sucrose hemolysis and acid-ham test. The patient was then started on prednisolone in a dose of 40mg per day. She was followed up with serial Hemoglobin and platelet count determination. This was her second pregnancy. The patient was booked at 8 weeks of gestation. She was subjected to the routine blood investigations and an ultrasound. Her hemoglobin at the first antenatal visit was 8mg% and platelets were 82,000/dl. She was advised to continue prednisolone. Hemoglobin and platelet counts were monitored every 2 weekly. The patient required blood transfusion 4 times in the antenatal period at 20, 24, 26 and 31 weeks of gestation.

We debated about starting the patient on anticoagulants but decided against it. The patient went into spontaneous labour at 32 weeks of gestation and did not respond to tocolysis. She delivered a 1.4 kilogram male baby. She received one unit packed cells. The patient developed fever on the third postpartum day which responded to antibiotics.

Discussion

Paroxysmal Nocturnal hemoglobinuria, a rare form of hemolytic anemia of unknown etiology is characterized by acute and chronic intravascular hemolysis with intermittent hemoglobinuria, severe anemia, infections and thrombosis of insidious onset. The episodes of PNH are triggered by stresses including stress of pregnancy.^[5] It is associated with a poor pregnancy outcome. Many patients are subfertile and may have spontaneous or missed abortions and pre-term deliveries. Anemia which is seen in PNH may be severe in pregnancy probably due to hemolytic crisis precipitated in pregnancy.^[6] The anemia may be

severe enough to warrant blood transfusion as seen in our patient who received blood transfusion 4 times during pregnancy.

Venous thrombosis is a common complication especially stimulated by pregnancy. Hepatic venous thrombosis and cerebral vein thrombosis are more sinister complications causing maternal deaths in the antenatal period or puerperium. Although anticoagulation is usually given to patients with current thrombosis or past history of thromboembolism^[7] some researchers have used anticoagulants prophylactically to prevent thrombosis especially in puerperium in women with PNH.^[6] Another main issue in PNH is thrombocytopenic bleeding. Serial platelet counts should be done during pregnancy and should be kept above 50,000/dl. Steroid therapy with prednisolone is the treatment of choice. However, the definite therapy is bone marrow transplantation.^[8]

The mode of delivery of these patients is usually vaginal, but caesarean may have to be done for obstetric indications in which case general anesthesia may be required,^[8] especially in patients with thrombocytopenia or on anticoagulants. Our patient was diagnosed early in pregnancy, was given prednisolone throughout pregnancy and required several blood transfusions for severe anemia. She had a spontaneous preterm vaginal delivery at 32 weeks gestation and gave birth to a healthy baby. As there was no past or present history of thrombosis, anticoagulation was not given. Our case highlights the importance of team effort of hematologist and obstetrician and strict vigilance in the form of regular antenatal monitoring, steroids and repeated blood transfusions for successful maternal and fetal outcome in patients with PNH.

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