Pregnancy in Budd Chiari Syndrome—A Case Report

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Introduction

BCS is a condition where there is obstruction to hepatic venous outflow due to thrombosis of the hepatic vein or inferior vena cava. The prevalence of BCS during pregnancy and puerperium ranges from 3.8 to 21.5% [1]. However, the incidence of pregnancy in a woman diagnosed with BCS is unknown due to its rarity. As BCS is commonly encountered in women of childbearing age group, pregnancy poses a challenge. Traditionally

pregnancy was discouraged in these women due to its prothrombotic state. However, with the increased use of anticoagulation and decompressive procedures, successful pregnancy outcomes have been observed. Despite management of this condition with a multidisciplinary team, worsening of the disease leading to hepatic failure requiring transplantation and case fatality has been encountered.

Case Report

A 21-year-old Gravida 1 with chronic BCS is presented to us with 9-week gestation. She was diagnosed with BCS 3 years ago as part of evaluation for menorrhagia and anemia. Blood investigations revealed pancytopenia. Her thrombophilia screen was negative. JAK2 and PNH (paroxysmal nocturnal hemoglobinuria) screens were negative. LFT was mildly deranged. An ultrasound examination revealed BCS with right hepatic vein obliteration with left hepatic vein obstruction with collaterals to capsular veins, splenomegaly and ascites. Upper GI endoscopy revealed early esophageal varices. Radiological attempt to open the hepatic vein was done but failed.

Three years later, she presented to us with pregnancy. She was started on Inj. Clexane 40 mg s/c OD since her first antenatal visit. Serial ultrasounds were done to monitor fetal growth and to detect ascites, a sign of worsening of the liver condition. Ultrasound image of Doppler study of the liver was as shown in the fig. (Ref. Figures 1, 2). Prophylactic corticosteroids were given at 30-week gestation in view of ultrasonography showing funneling of internal os and a cervical length of 2.7 mm. Patient had PROM at the 31-week gestation, liquor was stained with thick meconium, and the cervix was unfavorable for induction. NST was non-reactive. An emergency LSCS was performed in view of fetal distress. At the time of delivery, her Hb was 7.8 g/dl and platelet count was 51,000. Her liver function test revealed hypoalbuminemia, and the liver enzymes, however, were within normal limits. She received 3 units of platelet transfusion during the procedure. Intraoperatively, moderate amount of ascites, a well-formed lower uterine segment, and thick meconium-stained liquor were noted. A female baby weighing 1.3 kg with a placenta of 560 g (hyperplacentosis) was delivered.

Minivac wound drain was placed in the subcutaneous plane in view of moderate ascites. Postoperative management included 1 pint of packed RBC transfusion, fluid restriction, thromboprophylaxis with unfractionated heparin, diuretics and a high protein diet along with antibiotics. The surgical site was inspected and drain removed on the 7th postoperative day. Histopathology of the placenta showed extensive infarction. Heparin was replaced by warfarin and continued for 6 weeks postpartum. Minimal

Fig. 1 USG of liver showing non visualization of the hepatic vein proximally

Fig. 2 USG of liver showing occlusion of right hepatic vein

ascites was noted on the 12th postoperative day. The baby required NICU care for 3 weeks. Lactation was well established in the mother. Contraception was discussed, and the choice between intrauterine devices and barrier contraception was given. Patient is due to undergo a repeat radiological attempt at opening the hepatic vein at a later date.

Discussion

BCS is defined as obstruction of hepatic venous outflow anywhere between the small hepatic veins to the supra-hepatic inferior vena cava [2]. BCS mainly affects woman of the reproductive age group. With the prevalence of 1 in 100,000–500,000, primary BCS is rare disorder. The etiology and pathogenesis are not completely understood. BCS is primarily a vascular disorder and the presence of a

hypercoagulable state appears inevitable. Pregnancy being a hypercoagulable state with increase in serum clotting factors, BCS can develop during pregnancy. Prolonged bed rest and fluid restriction in the postnatal period which is believed to enhance the quality of breast milk in rural India leads to dehydration and increased clotting factors precipitating BCS, cerebral cortical vein thrombosis and DVT. In women already diagnosed to have BCS, pregnancy can lead to severe exacerbation of the disease. There is a risk of recurrence of hepatic vein thrombosis, ascites and can present with increasing abdominal distension, pain and dyspnea. Thrombocytopenia is frequently encountered due to hypersplenism. Maternal mortality can result from liver decompensation, bleeding varices, intraoperative and postoperative hemorrhage, deep venous thrombosis and pulmonary embolism. Despite this, a study by Rautou et al. [3] showed that in women with well-controlled BCS, pregnancy should not be contra indicated as maternal outcome and fetal outcome beyond gestation week 20 are good.

Impact of BCS on Pregnancy

Patients are bound to develop anemia due to the chronic liver condition. There is an increased risk of miscarriage, preterm delivery, IUGR, still births and pre-eclampsia. The risk of intrapartum and postpartum hemorrhage necessitating transfusions is increased as the patient is on anticoagulants. The need for thromboprophylaxis during pregnancy and the postpartum period and ICU care for the baby in view of prematurity and growth restriction increases the costs of treatment.

Impact of Pregnancy on BCS

There is an increased occurrence of bleeding esophageal varices, worsening ascites and thrombosis leading to

obstruction of venous shunts. Liver decompensation requiring liver transplantation has been noted. Merz et al. [4] recently reported a case of a pregnancy with BCS which ended lethally within 2 years of delivery despite liver transplantation.

Conclusion

Managing pregnancy with BCS is a challenge. Close monitoring, adequate anticoagulation, monitoring of fetal growth and the liver condition along with a multidisciplinary approach lead to successful outcomes.

Compliance with Ethical Standards

Conflict of interest The authors declare that there is no conflict of interest regarding the publication of this paper and they have not received any Grant.

Informed Consent Informed consent was obtained from the patient for presentation of the case report.

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