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CASE REPORT

Pregnancy Complicated by Portal Hypertension Secondary to Noncirrhotic Portal Fibrosis

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ABSTRACT

Noncirrhotic portal fibrosis (NCPF) is a liver disorder characterized by periportal fibrosis of small and medium branches of portal vein resulting in portal hypertension with the sequelae of variceal bleed requiring immediate attention. We report a case of 27-years-old primigravida with NCPF presented for her booking visit with 3 months of amenorrhea. Clinical examination and ultrasound revealed 12 weeks of intrauterine gestation with moderate splenomegaly, dilated portal vein, and splenic vein. The patient was referred to medical gastroenterologist. Hematological investigations were found normal except for borderline thrombocytopenia. Upper gastrointestinal endoscopy revealed grade I esophageal varices. She was monitored bimonthly by obstetrician and hepatologist as she was potential for variceal bleed, anemia, liver derangement, and coagulation abnormalities during antenatal and intrapartum period. She had well-preserved liver function throughout pregnancy and no variceal bleed. Fetal surveillance was done and she received prophylactic corticosteroids at 28 weeks of gestation for fetal lung maturation. Antenatal period was uneventful. In view of breech presentation, elective cesarean section was performed and a healthy male baby was delivered weighing 2.9 kg, with good Apgar score. Intrapartum and postpartum period was uneventful. Surveillance by a multidisciplinary team is important for such high-risk pregnancy for optimizing obstetric and neonatal care.

Keywords: Noncirrhotic portal fibrosis, Portal hypertension, Splenomegaly, Variceal bleed.

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INTRODUCTION

Noncirrhotic portal fibrosis (NCPF) is one of the important disease entities of the liver characterized by clinically significant portal hypertension with preserved liver function, splenomegaly, and absence of viral or other etiology of chronic liver diseases. Pregnancy with NCPF is uncommon. Management of NCPF in pregnant women is similar to that of nonpregnant. Prognosis depends on the severity of the disease and is usually good in this condition.

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CASE REPORT

A 25-years-old primigravida with 3 months amenorrhea reported for antenatal checkups. She was a known patient of NCPF for 2 years. She was apparently well 2 years back when she developed intermittent swelling of both lower limbs, painless progressive abdominal distension, and anemia for which she was investigated and found to have splenomegaly with free fluid in the peritoneum in the ultrasonography. Liver biopsy revealed focal mild portal fibrosis with no evidence of cirrhosis and grade I esophageal varices in upper gastrointestinal (GI) endoscopy. Patient was advised Tab. Propranolol 20 mg 1 od, Tab. Frusemide 20 mg 1 od, Tab. Spironolactone 50 mg 1 od, Tab. Ferrous sulfate 1 od, salt-free diet, 60 gm of protein, and fluid restriction and she was regularly followed up by the concerned specialist. Her vaccinations were up-to-date.

At the time of her first visit to the hospital, clinical examination revealed 12 weeks of gestation with splenomegaly which was enlarged to the level of the umbilicus, with no evidence of ascites or other features of chronic liver disease. Ultrasonographic study confirmed intrauterine gestation of 12 weeks and splenomegaly, dilated portal vein, splenic vein with multiple collaterals at hilum. She was referred to the medical gastroenterology clinic at the hospital to provide multidisciplinary team input to her care.

She was investigated and hematological tests were found normal except for borderline thrombocytopenia ($100,000/mm^3$). Upper GI endoscopy revealed small esophageal varices (grade I) with prominent vein in the fundus. All the drugs which she was consuming were stopped except iron tablets.

She had bimonthly antenatal visits with obstetrical and hepatology teams. During each visit, detailed clinical examination and routine investigations for maternal status and specific investigations for any

disease-related complications like variceal bleed, anemia, liver derangement, and coagulation abnormalities were performed. During her pregnancy, her platelets remained borderline (90,000–120,000/mm^3) and hemoglobin ranged from 9.0 gm/dL to 10.5 gm/dL. Her liver parameters and coagulation factors were normal. Significant concern remained about her potential for variceal bleed. Fetal anatomical scan was normal with a well-grown fetus of 50th centile. Due to the potential for preterm delivery, she received two doses of betamethasone at 28 weeks of gestation to aid lung maturation.

The pregnancy proceeded uneventful till term. The patient was taken up for elective cesarean section at term in view of breech presentation. A live male baby was delivered by breech extraction and transferred to the neonatologist. The baby weighed 2.9 kg with the APGAR score of 8 to 10. The remainder of the surgery was uneventful. Histology of the placenta was unremarkable. The patient had an uneventful postpartum course. She was advised contraception and underwent insertion of copper T 380A at 3 months postpartum.

DISCUSSION

Noncirrhotic portal fibrosis or idiopathic portal hypertension is a disease of uncertain etiology characterized by periportal fibrosis and involvement of small and medium branches of the portal vein, resulting in the development of portal hypertension with preserved liver function and structure.[1]

Pregnancy in a patient with portal hypertension presents a special challenge to the obstetrician as associated physiological hemodynamic changes worsen the portal hypertension. Risk of variceal bleed increases many fold during pregnancy which influences the maternal and perinatal outcome. Various studies have shown that the prognosis of portal hypertension due to NCPF is good when compared with portal hypertension due to cirrhosis. A state of mild, compensated, and disseminated intravascular coagulation secondary to endotoxemia or portosystemic collaterals has been reported in some cases of NCPF. Sarin and Kapoor[2] found derangements in the international normalized ratio and fibrinogen levels in their series of patients with NCPF. Conventional tests of liver function are normal or near normal.[3,4] Hypersplenism may occur in the antenatal period but usually asymptomatic.

All patients of moderate to massive splenomegaly with NCPF should have screening endoscopy.[1] Risk of variceal bleeding in pregnant women with portal hypertension is 19 to 45% due to increased portal pressures, reflux esophagitis, obstruction to the inferior vena cava by the gravid uterus. The timing and severity of hematemesis, however, are unpredictable. Portal hypertension due to NCPF patients tolerate variceal bleeding relatively well because of the well-preserved hepatic synthetic function.

Endoscopic variceal ligation (EVL) or sclerotherapy is effective in 80 to 90% of patients in controlling acute and rebleeding from esophageal varices[5,6] and can be performed safely during pregnancy. If varices are diagnosed prenatally, primary prophylaxis with EVL for large varices improves the pregnancy outcome. Repeat endoscopic evaluation is advised during pregnancy.

Anemia of microcytic, hypochromic (due to GI blood loss) or normocytic, normochromic, leukopenia (<4,000 mm^{-3}), and thrombocytopenia (platelets <50,000 mm^{-3}) may be present and are due to hypersplenism but usually asymptomatic. Jaundice, ascites, signs of chronic liver disease are uncommon. Liver function is well preserved in NCPF.

Frequent antenatal visits, and during each visit, complete physical examination, hematological tests, including liver function test, and coagulogram are done to know the status of the disease. Upper GI endoscopy should be performed in NCPF patients of moderate to massive splenomegaly and thereafter indicated. In our case, the patient was asymptomatic, hematological investigations were near normal, and upper GI endoscopy revealed grade I varices. Hence all the medications were stopped and monitored meticulously throughout pregnancy for the maternal and fetal conditions.

During intrapartum period, even though NCPF patients tolerate well and cesarean section is reserved for obstetrical indications, however there is risk of variceal rupture when the patient strains.[7] Sengstaken-Blackmore tube and cross-matched blood should be readily available and they are not allowed to bear down. To avoid straining, epidural analgesia is ideal with elective forceps or ventouse extraction in the second stage of labor if they are given a trial. In our case, in view of breech presentation, she underwent elective cesarean section uneventfully. Temporary contraception with intrauterine device is preferred.

CONCLUSION

Pregnancy is not contraindicated in patients with portal hypertension due to NCPF and the maternal prognosis is usually good. Termination of pregnancy needs to be considered only in patients with recurrent hematemesis and deranged liver function, especially abnormal coagulation profile. Management of pregnancy should be done by a multidisciplinary approach at tertiary care centers with backup facilities for intensive care and blood transfusion.

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