Pregnancy with portal hypertension

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Introduction:

Pregnancy with portal hypertension is an uncommon condition. A number of patients with Extra hepatic portal venous obstruction (EHPVO) and non cirrhotic portal fibrosis (NCPF) are surviving to adult life. In patients with cirrhosis, as long as liver function is relatively preserved, pregnancy is possible. Obstetricians will therefore, see more patients with portal hypertension in the future. They should be aware of the course of the disease during pregnancy and the management strategies to be adopted.

Case Report

A 23-year-old primigravida, who was a known patient of NCPF, reported for antenatal care at 24 weeks of gestation with complaints of a dragging sensation in the left upper abdomen. There was no history of fever, jaundice, hematemesis, bleeding tendencies or blood transfusion in the past. Examination revealed pallor, uterine enlargement commensurate with the period of gestation, hepatomegaly and massive splenomegaly. There were no stigmata of chronic liver disease. Liver function and coagulation parameters were normal. Markers of infective hepatitis were negative. Pancytopenia was present and attributed to hypersplenism. Sonographic examination confirmed hepatomegaly without distortion of liver architecture, massive splenomegaly, dilatation of portal collaterals, absence of ascitis and a single, live intrauterine fetus corresponding to the period of gestation. Upper gastrointestinal endoscopy was done and it revealed the presence of grade III varices.

The patient was hospitalized and given tablet propranolol 20 mg three times a day and hematinics. The pregnancy proceeded uneventfully. The patient was taken up for elective LSCS at 38 weeks duration and a live male neonate weighing 2.1 kg delivered. The liver was inspected at surgery and appeared normal externally. There was no ascitis. Postoperative recovery was uneventful. The patient underwent splenectomy and shunt surgery six months post partum.

Discussion

Prognosis of portal hypertension during pregnancy depends upon the underlying cause and the extent of derangement of liver function. Maternal prognosis is better with EHPVO and NCPF and poor with cirrhosis of the liver. Maternal mortality ranges between 2% and 18%; being maximum with cirrhosis. The causes of death are generally hematemesis, hepatic coma or postpartum hemorrhage. Perinatal mortality ranges between 11% and 18%, mainly due to preterm delivery or intrauterine growth restriction (IUGR).

Of the women with cirrhosis, 20-30% will have hematemesis during pregnancy with the mortality ranging between 50-60%. The incidence of hematemesis in patients with EHPVO and NCPF is around 7%. The timing and severity of hematemesis, however are unpredictable. Hematemesis is more common in pregnancies complicated by varices. Hematemesis during pregnancy is contributed to by increased portal pressures during pregnancy, reflux esophagitis and obstruction to the inferior vena cava by the gravid uterus.

Management of portal hypertension in pregnant women is similar to that in non-pregnant patients. Beta blockers are given to reduce portal venous pressures. Surgical management by banding and sclerotherapy have been successfully employed during pregnancy. It is possible to do shunt surgery during the second trimester.
There is a danger of variceal rupture and hematemesis when the patient strains during labor. Patients with EHPVO and NCPF generally tolerate labor well and cesarean section is not mandatory. It is however, necessary to have a Sengstaken-Blackmore tube and adequate amount of cross matched blood readily available if these patients are given a trial of labor. They must not be allowed to bear down and the second stage should be cut short.

Pregnancy is not contraindicated in patients with portal hypertension due to NCPF, EPVOC and compensated cirrhosis. Termination of pregnancy needs to be considered only in patients with decompensated cirrhosis, recurrent hematemesis and deranged liver functions, especially abnormal coagulation profiles. The management of pregnancy with portal hypertension should only be done at tertiary care centres by a multidisciplinary team with backup facilities for intensive care and blood transfusion.

References