A case of extra hepatic portal vein obstruction in pregnancy with superimposed pre-eclampsia

Bindu Nambisan*, Sreekumary Radha, Mayadevi Brahmanandan, Libu Gnanaseelan Kanakamma

Department of Obstetrics and Gynaecology, Medical College, Trivandrum, Kerala, India

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*Correspondence:
Dr. Bindu Nambisan,
E-mail: bindu.nambisan1971@gmail.com

ABSTRACT

Extra hepatic portal vein obstruction in pregnancy poses a clinical challenge by itself. We present here a case of a 19 year old primigravida with EHPVO who developed superimposed preeclampsia. She had a successful maternal and fetal outcome in a tertiary care centre owing to the team effort involving specialists from medical gastroenterology, nephrology, anesthesiology, and neonatology apart from senior obstetrician. EHPVO is an important cause of non-cirrhotic portal hypertension in third world countries. In pregnancy, the increased blood volume and cardiac output and mesenteric vasodilatation will increase portal flow and aggravate portal hypertension in these patients. The resultant haematemesis in such patients, can compromise the perinatal outcome. Endoscopic variceal ligation (EVL) reduces the risk of variceal bleeding, and can improve the pregnancy outcome in these women. Thrombocytopenia due to splenomegaly is one of the major complications in these patients and has to be corrected before pregnancy. Platelet transfusion is required intrapartum if the count is less than 50,000/mm³ caesarean delivery is reserved only for obstetric indications.

Keywords: EHPVO, Preeclampsia, EVL

INTRODUCTION

Extra hepatic portal-vein obstruction (EHPVO) is defined as obstruction of the extra hepatic portal vein with or without the involvement of the intrahepatic portal veins or splenic or superior mesenteric veins. In developing countries, it is a common cause of portal hypertension, accounting for up to 30% of all variceal bleeders. EHPVO is an important cause of non-cirrhotic portal hypertension in third world countries. The most common site of block is at the portal vein formation (90%) and total block of splenoportal axis is seen in 10% of cases. The etiology and clinical features are different in children and adults. In children, the causes are umbilical sepsis, neonatal systemic sepsis, umbilical catheterization and developmental anomalies. Other causes include dehydration, multiple exchange transfusions and sepsis.

In adults, important causes are neoplastic diseases, infections, pancreatitis, myeloproliferative disorders and hypercoagulable states. The cause of portal vein block is obscure in 50% of cases. In pregnancy, the increased blood volume and cardiac output and mesenteric vasodilatation will increase portal flow and aggravate portal hypertension in these patients. The resultant haematemesis in such patients, can compromise the perinatal outcome. Prenatal obliteration of high-risk varices by endoscopic sclerotherapy (EST) or endoscopic variceal ligation (EVL) reduces the risk of variceal bleeding, and can improve the pregnancy outcome in these women. In the Western countries, portal hypertension is usually due to cirrhosis. Also, these women with EHPVO have normal fertility, unlike women with cirrhosis who have reduced fertility and up to 40% fetal-loss rate. Increasing intra-abdominal pressure in the
second and third trimesters also contributes to portal hypertension by increasing the inferior vena cava pressure.\(^8\) This results in rerouting of blood via gastroesophageal collaterals and increases the risk of variceal bleeding. The presentation could be either acute (recent) or chronic EHPVO. Patients with acute EHPVO usually present with abdominal pain, ascites, jaundice, or fever.\(^2\) The majority of patients with chronic EHPVO present with repeated bleeding episodes from esophageal varices. The incidence of variceal bleeding in pregnancy in patients with EHPVO has been reported to range from 20% to 34%.\(^5\) However, it is generally agreed that patients with a prenatal diagnosis of EHPVO have a much lower incidence of variceal bleeding compared to those who are diagnosed during pregnancy.\(^6\) Primary prophylaxis by EVL is recommended for high-risk varices before pregnancy. Either beta-blockers or endoscopic therapy can be used for primary prophylaxis of variceal bleeding in patients not planning pregnancy.\(^2,10\)

Beta-blockers reduce the portal pressure by reducing the cardiac output and by causing splanchnic vasoconstriction.\(^9\) However, if used during pregnancy, they can cause fetal bradycardia and growth retardation. Thrombocytopenia due to splenomegaly is one of the major complications in these patients and has to be corrected before pregnancy. Platelet transfusion is required intrapartum if the count is less than 50,000/mm\(^3\).\(^7\)

The role of anticoagulant therapy in chronic EHPVO is not clear. However, in patients with documented prothrombotic disorders, lifelong thromboprophylaxis is recommended.\(^1\) Repeat endoscopic evaluation is usually done during pregnancy, and either EST or EVL may be used to obliterate the residual varices.

Caesarean delivery is reserved only for obstetric indications. The delivery should be monitored closely, and second stage of labour may be cut short by operative vaginal delivery in patients who are at risk.\(^2\) Intravenous fluids should not be administered overzealously because of the risk of volume overload and variceal bleeding.

**CASE REPORT**

Mrs A, 19 years old primigravida presented at 32 weeks of gestation with swelling of both legs and severe vulval oedema since past 2 days. On admission she had difficulty in walking due to vulval oedema. Married since 2 years and it was a spontaneous conception. Had regular antenatal checkups from our hospital. She was a known case of EHPVO on treatment from medical gastro since the age of three which was diagnosed when she presented with haemetemesis during childhood. She was asymptomatic since then but on follow up. During the 3rd month of pregnancy she had a bout of haemetemesis and an endoscopic varicose ligation was done and 5 bands applied. She had so far no history of hypertension but on admission was detected to have a BP of 150/100 mmHg not associated with any headache, blurring of vision, or epigastric pain. Her BRE, URE, LFT, RFT and platelet count was sent the same day and the initial report revealed 3+ proteinuria, with a marginally raised creatinine and a low platelet count and normal liver enzymes. A medical gastro opinion was sought on the same day and rtp a liver function test, peripheral smear and ultrasound scan was planned. Nephro opinion was that it was a case of preeclampsia induced renal dysfunction. By 12 hours after admission there was severe oliguria and termination of pregnancy was planned taking into consideration renal dysfunction. USS revealed an IUGR foetus of growth corresponding to 29 weeks with adequate liquor with moderate ascites and B/I hyperechoic maternal kidneys. LSCS was done under epidural anaesthesia. A preterm baby of weight 1 kg was delivered. Grade 1 abruption was also noted and there was about 1.5 liters of ascitic fluid. Post procedure patient was shifted to intensive care for close monitoring. She was put on broad spectrum antibiotics and antihypertensives. As per nephrologists advice potassium was restricted. By the third postoperative day UOP showed significant improvement and BP was well controlled. Her liver and renal parameters were back to normal by about 1 week. However she continued to be in the hospital as baby was under the care of neonatologist and both mother and baby were discharged in good general condition by 25th postoperative day with a baby weight of 1.3 kg.

**DISCUSSION**

This case demonstrates how a team effort involving multidisciplinary approach in a tertiary level institute can change the prognosis in clinically challenging cases. Here a team effort involving medical gastro, nephro, anaesthesia and neonatologist along with obstetrician skilled in handling high risk cases has made a difference and both mother and baby were discharged in good general condition. This case also brings forth the diagnostic dilemmas involved when a condition of preeclampsia is superimposed on a patient with EHPVO.

In conclusion, women with EHPVO would have a good pregnancy outcome if they were managed in a tertiary care centre with a multidisciplinary approach.

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