PORTAL HYPERTENSION IN PREGNANCY - COMPLICATED BY HYPERSPLENISM AND SEVERE THROMBOCYTOPENIA WITH SUCCESSFUL OBSTETRIC OUTCOME

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Abstract

Pregnancy is a rare event in patients with portal hypertension, carrying with a host of maternal and foetal complications. Judicious decision regarding continuation or termination of pregnancy has to be taken, after considering the patient’s desire for childbearing & the severity of underlying disease process, as also the necessary facilities and expertise to manage accompanying complications.

Keywords: Portal hypertension, Hepatosplenomegaly, Pancytopenia

Introduction

Pregnancy with portal hypertension is rare but worrying for the clinician. Investigation and management of portal hypertension before and at the beginning of pregnancy can reduce the risks of foetal loss, restricted intra-uterine growth, premature birth and maternal mortality, which are closely related to gastrointestinal haemorrhage. The risks related to the underlying disease, such as liver failure with cirrhosis and thromboembolic risk with vascular diseases associated with thrombophilia must be taken into consideration. Generally, vaginal delivery with early analgesics for the mother assisted by an extraction device should be preferred to caesarean section, which must be reserved for obstetrical indications.

Case Report

A 21-year-old primigravida reported for antenatal checkup to the general OPD of ESI-PGIMSR, Joka, Kolkata at 12 weeks of pregnancy. She complained of frequent bleeding from gums besides the usual early pregnancy symptoms. At 24 weeks of gestation, patient complained of melena.

On general examination, there was pallor, jaundice, mild ascites, moderate hepatomegaly and massive splenomegaly (20cm) reaching below the umbilicus, and uterine enlargement commensurate with period of amenorrhoea. Liver function tests and coagulation profile were normal. Markers of infective hepatitis were negative. Haemogram revealed pancytopenia, especially progressive & severe thrombocytopenia.

Sonographic examination at 24 weeks confirmed hepatomegaly, without distortion of liver architecture, massive splenomegaly with dilated splenic vein, dilatation of portal collaterals and mild ascites (Figures 1, 2 & 3). Gall bladder showed calculus. There was a single live intrauterine foetus. Foetal growth was adequate for the period of gestation.

Figure 1. Dilated portal vein
Figure 2. Spleen enlarged to 21.5 cm

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Upper gastroendoscopy, done at 24 weeks, revealed dilated, bleeding oesophageal varices, thus banding was performed at the same sitting (Figure 4).

Patient was hospitalized at 30 weeks gestation due to severe thrombocytopenia and frequent platelet transfusions had to be given. A total of 64 units were given antepartum. Moderate amount of ascitic fluid accumulated, splenomegaly increased and frequent bouts of melena were reported as pregnancy progressed. Clinical and sonographic follow up of the foetus, including Doppler study showed adequate foetal growth for that gestational age. At 38 weeks and 3 days, patient went into spontaneous labour; cardiotocography indicated foetal distress and baby (with birth weight 2.4 kg and good apgar score) was delivered by caesarean section. Postpartum bleeding was within normal limits. During the postpartum period, platelet count remained low and 8 units of platelet transfusion had to be given in first postpartum week. After puerperal period, patient was temporarily lost to follow up.

Due to dragging pain and severe fatigue, patient reported to medical OPD after 9 months following childbirth. Sonographic examination revealed splenomegaly of 21 cm. Haemogram showed moderate pancytopenia. Decision for splenectomy was taken, but prior to that, liver biopsy was done.

Liver biopsy revealed liver parenchyma with focal sinusoidal dilatation and prominent ballooning degeneration of hepatocytes. Portal tracts were histologically normal. Patient underwent banding (3 bands) of the grade 2 oesophageal varices followed by splenectomy. Post splenectomy, patient had an uneventful recovery. At present, patient is being followed up for further variceal bleeding and her haemogram is showing normal RBC, WBC and platelet count.

Histopathology of spleen revealed extensively congested and dilated splenic sinusoids with focal parenchymal fibrosis and splenic vascular thrombosis (fibrocongestive splenomegaly).

Discussion

Portal hypertension (PH) is a clinical syndrome, defined as the elevation of hepatic venous pressure gradient (HPVG) above 1,2,3,4 mm of Hg. The HVPG measurement has been accepted as the gold standard for assessing the severity of portal hypertension, and has replaced the old one i.e. contrast angiography. The PH is considered to be clinically significant when HVPG exceeds 10 to 12 mm Hg, since this is the threshold for the clinical complications of PH to appear. PH is caused by a combination of two haemodynamic processes, occurring simultaneously:-

1. Increased intrahepatic resistance to blood flow through the liver due to cirrhosis
2. Increased splanchic blood flow secondary to vasodilatation within the splanchic vascular bed.

The most common cause of portal hypertension is cirrhosis (90%) or scarring of liver. The pathological hallmark of cirrhosis is the development of scar tissue that replaces normal parenchyma, blocking the portal flow of blood through the organ and disturbing normal hepatic function. The gold standard for diagnosis of cirrhosis is a liver biopsy. The fibrous tissue bands (septa) separate hepatocyte nodules. Eventually, the septae replace the entire liver architecture, leading to decreased blood flow throughout the liver. The backflow of blood leads to congestive splenomegaly, hypersplenism and increased sequestration of platelets. Portal hypertension is responsible for most of the complications of cirrhosis. Our patient had severe thrombocytopenia during pregnancy, thus liver biopsy was not undertaken during gestation, fearing the risk of haemorrhage.
Portal hypertension may also be caused by:-

(a) Thrombosis in the portal vein-Extra hepatic portal venous obstruction (EHPVO)

(b) Noncirrhotic portal fibrosis (NCPF) with portal venous obliteration and intralobular slender fibrosis.

Collaterals exist between the portal and systemic veins. The resistance in the portal vessels is normally lower than in the collateral circulation, and blood flows from the systemic bed into the portal bed. However, when PH develops, the portal venous pressure is higher than systemic venous pressure, and this leads to reversal of flow in these collaterals (i.e. from portal to systemic veins). In addition, the collateral circulatory bed develops new fragile blood vessels (varices) across the oesophagus and stomach in an attempt to decompress the portal circulation. These new collaterals are fragile and insufficient to decompress the portal circulation. The raised portal pressure causes variceal rupture and bleeding. Oesophageal variceal bleeding has been reported in 18-32% of pregnant women with cirrhosis and 50% in those with associated portal hypertension with mortality varying from 18-50%. In contrast, noncirrhotic portal hypertension with pregnancy fares better with 2%-6% incidence of haematemesis, perhaps because cirrhosis is more commonly associated with deranged coagulopathy.

Ascitis develops insidiously in cirrhotic portal hypertension (75%) and dyspnoea occurs due to abdominal distension and/or accompanying pleural effusion. Ascitis may be complicated by infection, especially spontaneous bacterial peritonitis (SBP). The most important analysis of ascitic fluid are cell count, fluid culture, calculation of Serum-ascites albumin gradient (SAAG) which correlates with portal venous pressure. If SAAG is >/= 1.1g/dl, ascites is ascribed to portal hypertension with approximate 97% accuracy. Our patient developed ascites from sixth month of pregnancy.

Hypersplenism commonly goes hand in hand with massive congestive splenomegaly in patients with cirrhosis and portal hypertension. Hypersplenism is associated with thrombocytopenia, leucopenia, anaemia or a combination of any of the three. Severe thrombocytopenia may increase the risk of bleeding. The four cardinal signs of hypersplenism are (1) Splenomegaly, (2) Reduction in the number of circulating blood cells affecting granulocytes, erythrocytes or platelets in any combination, (3) A compensatory proliferative response in the bone marrow (4) Potential for correction of these abnormalities by splenectomy.

Splenomegaly is usually associated with increased workload (such as in hemolytic anaemia), which suggests that it is a response to hyperfunction. An enlarged spleen, along with caput medusa, is an important sign of portal hypertension. Our patient had massive splenomegaly, which was detected from 20 weeks of gestation and which persisted even after childbirth. She also had severe and persistent thrombocytopenia and anaemia throughout her pregnancy, puerperium and 1.5 years after childbirth. She underwent splenectomy following which there was no further pancytopenia. Thus, she satisfied 3 of the 4 cardinal points of hypersplenism; response of bone marrow to pancytopenia was not documented since bone marrow biopsy was not done.

Variceal bleeding occurs commonly during second and third trimester when maternal blood volume is maximally expanded. There is increased portal pressure, reflux oesophagitis and obstruction to the inferior vena cava by the gravid uterus. Variceal haemorrhage may be in the form of haematemesis &/or malena. Our patient had malena from the fourth month of gestation till the end of pregnancy.

Portal hypertension and cirrhosis can cause life-threatening variceal haemorrhage, hepatic decompensation, splenic artery aneurysm and rupture as well as postpartum haemorrhage. Postpartum uterine haemorrhage may occur in 7% to 10% of pregnancies in patients with cirrhosis. This is possibly due to coagulopathy and thrombocytopenia and may be as high as 18% versus 1.08% in the noncirrhotic population.

Foetal Outcomes- The spontaneous abortion rate in patients with cirrhosis is significantly higher (30%-40%) than that of the general population (15% to 20%). Perinatal mortality (11% and 18%) is due to preterm delivery (spontaneous or iatrogenic) or intrauterine growth restriction. Termination of pregnancy most often occurs as a result of variceal hemorrhage, stillbirth, intrauterine growth retardation, and maternal complications during antenatal period like pancytopenia/thrombocytopenia.

Management of portal hypertension in pregnant women is similar to that in non-pregnant patients. Beta blockers or nitrates are given to reduce portal venous pressure. Surgical treatment by banding and sclerotherapy has been successfully employed during pregnancy, especially as an emergency procedure for acute haemorrhagic episode. In banding, a gastroenterologist uses rubber bands to block the blood supply to each varix. In sclerotherapy, the gastroenterologist injects a solution into the bleeding varices to cause sclerosis, thereby arresting the bleeding. Our patient had frequent bouts of malena during pregnancy for which she underwent oesophageal variceal banding from 20 weeks of pregnancy.

If banding fails to control the variceal bleeding, one of the decompression procedures may be undertaken safely in the second trimester: Transjugular Intrahepatic Portosystemic Shunt (TIPS) or Distal Splenorenal Shunt (DSRS).

In the TIPS procedure, a tunnel is created within the liver, a stent is placed in it which connects the portal vein to one of the
hepatic veins, thereby rerouting blood flow in the liver and reducing pressure in the varices. In the DSRS procedure, the splenic vein is detached from the portal vein and connected to the left renal vein, thus bypassing the splenic vein. Hypersplenism causing severe thrombocytopenia requires splenectomy. However, portal hypertension will persist if the pathophysiology of the disease remains uncorrected and repeated banding will be needed to manage bleeding varices.

Pregnancy may be allowed to go to term in tertiary care centres if coagulation profile and platelet count is maintained and blood transfusion facilities are adequate. Caesarean section is not mandatory in patients with EHPVO & NCPF since they tolerate labour well. Vaginal delivery may be allowed if obstetric factors are favourable. However, there is a danger of variceal rupture and haematemesis when the patient strains during labour or carries out valsalva manoevers. Second stage of labour must be cut short by episiotomy and extraction devices. Adequate blood must be kept ready to combat postpartum haemorrhage.

Conclusion

Pregnancy is not a contraindication in patients with portal hypertension due to NCPF, EPVOC and compensated cirrhosis. Pregnancy termination may be considered only in cases of recurrent haematemesis, decompensated cirrhosis, deranged liver functions and abnormal coagulation profiles. However, 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.

Editor's comment

The presented case highlights the importance of multidisciplinary approach when managing complex conditions during pregnancy. Pregnancies with portal hypertension are best managed in a tertiary level facility with access to surgical, radiological, perioperative, and medical specialists.

References