Case report

Pregnancy with Non-cirrhotic Portal Hypertension: A Case Report

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Abstract:
Pregnancy with non cirrhotic portal hypertension is an uncommon condition. Due to physiological changes in pregnancy, the patients with portal hypertension may exaggerate and produce life threatening complicationslike variceal haemorrhage and increased incidents of adverse maternal and fetal outcomes. Pregnancy also predisposes the patient to develop hepatic decompensation. So, management of such cases requires a multi-disciplinary approach. In this case report,21-year-old primigravida presented with known case of non cirrhotic portal hypertension at her 33 weeks of pregnancy with huge splenomegaly and history of hematemesis.The pregnancy ran uneventfully, and she underwent elective LSCS at 38+ weeks and gave birth of a live female baby, weighing 3kg was delivered. Post operative recovery was uneventful. The patient was discharged on the 4th post operative day. We want to focus upon different aspects of pregnancy with portal hypertensionto prevent complications and manage the case.

Keywords: Primigravida, Non-cirrhotic portal hypertension

Introduction:

Pregnancy is a state wherevarious anatomical, physiological & biochemical changes occur progressively. These changes may create a problem in the presence of portal hypertension. Non cirrhotic portal hypertension refers to a heterogenous group of liver disorders that primarily affect the liver vascular system that are classified anatomically blood flow as prehepatic, hepatic and post hepatic.[1,2]

In developed countries, cirrhosis of liver is the most common cause of portal hypertension. In developing countries extrahepatic portal vein obstruction contributes significantly to non-cirrhotic portal hypertension. The hepatic synthetic function is relatively well preserved and so is fertility[3,4] pregnancy in a patient with portal hypertension is a unique problem that needs specialized care to prevent potentially life-threatening complications such as gastrointestinal haemorrhage it is therefore important to understand the effect of pregnancy on portal hypertension and vice versa so that untoward incidents like fetal morbidity, mortality and gastrointestinal haemorrhage can be avoided.

Case study:

A 21-year-old primigravida report for antenatal care at 33 weeks of gestation with known case of non cirrhotic portal hypertension. Her LMP was on 08/02/23, accordingly EDD was on 15/11/23. She gave history of haematemesis six months back followed by hospital admission from 07/03/23 to 11/03/23 under supervision of a hepatologist. She gave history of 3-units blood transfusion due to massive hemorrhageat that time. From then she was gettingTab. carvedilol 6.25mg.

Examination revealed mild degree anaemia, uterine enlargement commensurate with period of gestation and huge splenomegaly. There is no stigmata of chronic liver discase. The liverfunction test and coagulation profile were normal. Viral markers were negative. Total count of WBC was 4.8 µ/L, Platelet 75µ/L.

Doppler study of upper abdomen revealed chronic liver disease features and normal doppler findings. USG of upper abdomen: features of chronic liver

disease with portal hypertension. Patient was advised for EVL after delivery. The pregnancy run uneventfully, and she underwent elective LSCS at 38 + weeks and gave birth of a live female baby, weighing 3kg was delivered. The liver was inspected per-operatively and appeared normal. There was no ascites. Post operative recovery was uneventful. The patient was discharged on 4th post operative day.

Discussion:

In pregnant women, cirrhosis is uncommon while viral or autoimmune related cirrhosis is more common in developing countries. The non-cirrhotic causes of portal hypertension include extra-hepatic portal vein obstruction, non cirrhotic portalfibrosis, portal vein thrombosis, Budd-Chiari syndrome,infection or congenital hepatic fibrosis.[5]

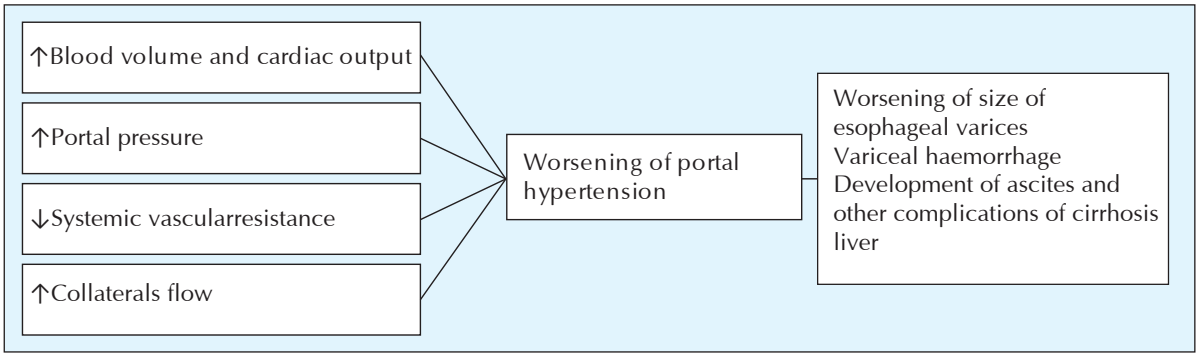

Figure-1: Effect of pregnancy hemodynamics on portal hypertension[5]

Pre-sinusoidal	Sinusoidal	Post-sinusoidal
● Portal vein obstruction ● Porto-sinusoidal vascular disease (PSVD) ● Schistosomiasis ● Arteriovenous fistulas ● Polycystic liver disease ● Congenital hepatic fibrosis ● Biliary diseases (primary biliary cirrhosis. primary sclerosing cholangitis)	● Drug-induced ● Acute fatty liver of pregnancy ● Alcoholic liver damage ● Non-alcoholic steatohepatitis ● Viral hepatitis ● Amyloidosis ● Infiltrative diseases ● Gaucher's disease ● Visceral leishmaniasis	● Budd-Chiari syndrome ● Veno-occlusive disease ● Primary vascular malignancies ● Hypervitaminosis A ● Epithelioid hemangioendothelioma and angiosarcoma

Figure-2: Etiologies of non-cirrhotic portal hypertension (NCPH)[6]

The prognosis of portal hypertension during pregnancy depends upon the underlying cause and the extent of derangement of liver function. Maternal prognosis is better with noncirrhotic portal hypertension 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. The mother is also at risk of developing severe anemia, splenic artery aneurysm rupture, ascites, spontaneous bacterial peritonitis. Perinatal mortality ranges between 11% and 18%, mainly due to preterm delivery or intrauterine growth restriction (IUGR).[5]

Since variceal bleed is the single important complication linked with poor pregnancy outcome, the basic aim is to prevent it. Endoscopy is the gold standard to assess the risk of bleeding in patients with esophageal varices. Upper gastrointestinal endoscopy is safe during pregnancy, the main risk being fetal hypoxia due to sedation or positioning.[5] Current American Association for the Study of Liver Disease (AASLD) recommendations include screening endoscopy in the second trimester as that is the time of maximum increase in the portal pressure.[9]

The treatment options in presence of esophageal varices are both medical and surgical. Non-selective beta blockers used to reduce portal pressure also reduce the risk of first bleed by half but the principal risk of using them in pregnancy is fetal growth restriction and fetal bradycardia Endoscopic variceal ligation of the large EV varices can also be done during pregnancy to prevent variceal bleeding. Current literature (Baveno V

consensus workshop) recommends EVL for acute esophageal variceal bleed, although, endoscopic sclerotherapy may be used if banding is technically difficult.[10]

Pregnancy can be allowed to go to term if the disease is well compensated. Early termination of pregnancy may be warranted in case of any obstetrical indication or progressive liver failure. In case of planned termination before 34 weeks, antenatal corticosteroids can be administered for fetal lung maturity. There are no recommendations as to the preferred mode of delivery- vaginal versus caesarean section in patients with portal hypertension.

The management during labor needs to be individualized depending on cause of portal hypertension and the disease status. Adequate amount of blood and plasma should be arranged and measures for balloon tamponade for the variceal hemorrhage must be handy. The second stage of labor may be shortened prophylactically to avoid overstraining by the mother.[11] The third stage should be managed actively, methergine should be avoided amongst the oxytocins. Postpartum hemorrhage should be anticipated and managed vigilantly.[12]

Conclusion:

Counseling prior to conception, regular follow-up and monitoring are essential in patients with portal hypertension. Pregnant women with NCPH should be managed at a tertiary care level with a multidisciplinary team including an obstetrician, hepatologist, anesthesiologist, and perinatologist. There are currently no clear protocols for treating NCPH during pregnancy, which raises the risk of negative outcomes for both the mother and the fetus. Further studies are also required to analyze more cases to evaluate the feasibility of different methods of pregnancy termination in patients with NCPH and how to deal with the maternal and perinatal outcomes.

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