Idiopathic Chronic Liver Disease in Pregnancy with a Complex Interplay of Portal Hypertension and Oesophageal Varices – A Challenging Clinical Conundrum

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ABSTRACT

The occurrence of idiopathic chronic liver disease in pregnancy, complicated by portal hypertension and oesophageal varices, presents a rare and high-risk obstetric challenge. This case highlights a successful multidisciplinary approach for managing a 23-year-old pregnant woman diagnosed with extrahepatic portal venous obstruction and Grade III oesophageal varices. Optimal coordination among obstetricians, medical gastroenterologists, and hematologists resulted in favorable maternal and neonatal outcomes. This case emphasizes the significance of early diagnosis, surveillance, and individualized delivery planning in women with hepatic vascular disorders during pregnancy.

Keywords: **Extrahepatic portal venous hypertension, Oesophageal varices, Splenomegaly, Thrombocytopenia, High-risk pregnancy**

INTRODUCTION:

Portal hypertension in pregnancy is rare, occurring in approximately 1 in 10,000 to 1 in 50,000 pregnancies. Among women of reproductive age, up to 15% may present with extrahepatic portal venous hypertension without cirrhosis. Pregnancy-induced hemodynamic changes, including a 45% increase in plasma volume, can exacerbate portal pressure and increase the risk of variceal bleeding. Around 30–50% of these women experience complications such as variceal hemorrhage, hepatic decompensation, ascites, splenomegaly, thrombocytopenia, jaundice, and fetal complications like preterm labor and fetal growth restriction.

AIM

To discuss the clinical approach and multidisciplinary management of pregnancy in a woman with idiopathic chronic liver disease and extrahepatic portal venous obstruction complicated by oesophageal varices.

CASE REPORT

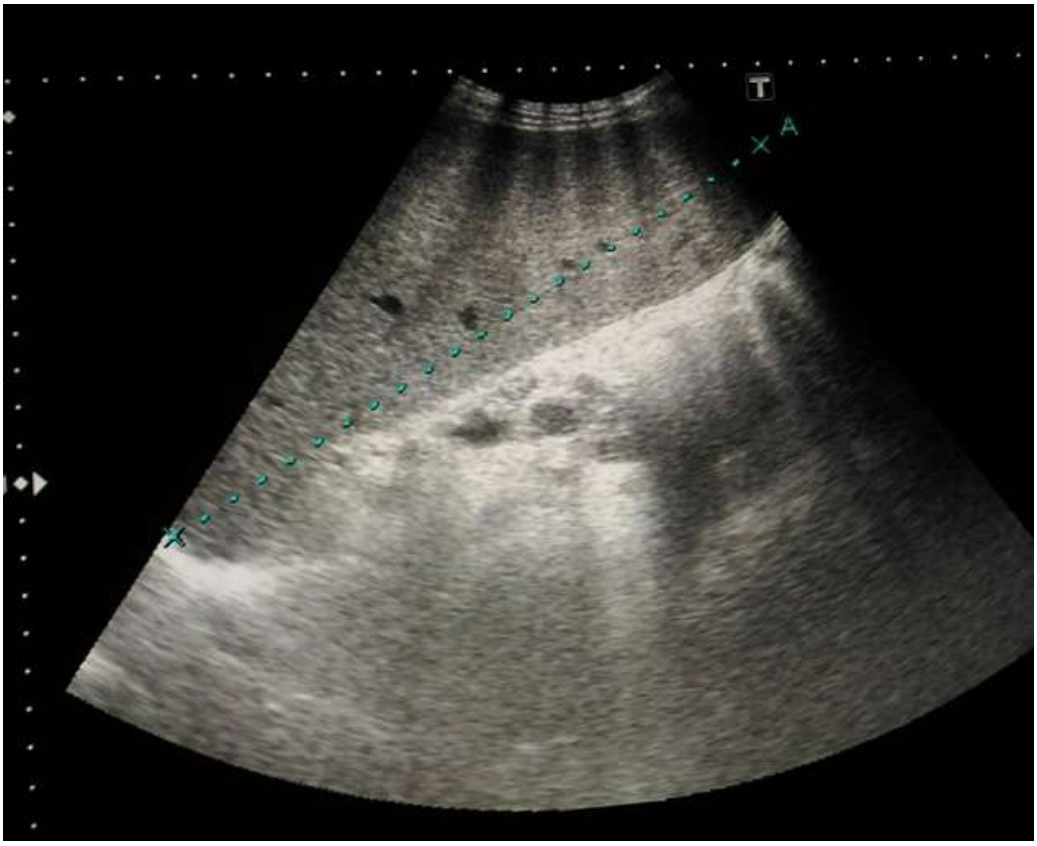
A 23- year- old Booked, G3P1L1A1 woman at 38 weeks+5days, a known case of Idiopathic chronic liver disease with Extrahepatic portal venous obstruction diagnosed 5 years ago and evaluated further with CECT abdomen and pelvis showing moderate splenomegaly with a splenic index of 3600 and Upper GI Endoscopy showing esophageal varices-Grade III for which she has underwent endoscopic variceal ligation after her 1st delivery, presented to SRIHER with leaking per vagina. She was found to have pre-labor rupture of membranes. She had moderate anemia, was on oral iron twice daily. She had a history of admission at 34 weeks+3days in view of threatened preterm labor and steroids were covered.

1- unit PRBC transfusion was also done at 34 weeks for Hb<8g/dl. MRCP done in 2nd trimester revealed portosystemic collaterals with cavernous transformation of portal vein. History of 1 Medical termination of pregnancy at 14 weeks was done in view of maternal condition, D&C done. She had regular antenatal checkups. Medical Gastroenterology opinion was sought, advised to taper Tab. Carvedilol 3.125mg to once daily and to watch for bradycardia, to cut short second stage of labor as the patient carries moderate risk of decompensation with a MELD score of 7. Hematologist opinion was obtained for thrombocytopenia and anemia, advised to transfuse 4 units FFP followed by 4 units RDP/1unit SDP, the same followed.

On admission, Hb-8g/dl, Platelet count was 30,000. Patient had persistent thrombocytopenia and normocytic hypochromic anemia throughout her pregnancy. On examination, she was hemodynamically stable with pallor and mild pedal edema. Abdominal examination revealed mild hepatosplenomegaly without ascites. Obstetric examination revealed a viable intrauterine pregnancy with normal fetal growth parameters.

Patient had a spontaneous vaginal delivery with episiotomy. Puerperal period was uneventful. Repeat platelet count after transfusion was 67,000 after delivery. She was also counselled for interval sterilisation and advised barrier contraception till then.

Figure 1: Abdominal ultrasonography shows coarse echotexture of the liver with increased portal vein diameter suggestive of chronic liver disease.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DISCUSSION

Chronic liver disease in pregnancy, especially of idiopathic origin, presents significant diagnostic and management challenges. Portal hypertension in such cases may manifest with variceal formation, posing a high risk of gastrointestinal bleeding. The Non-cirrhotic causes of portal hypertension include extra-hepatic portal vein obstruction, Non-cirrhotic portal fibrosis, Portal vein thrombosis, Budd-Chiari syndrome, infection or congenital hepatic fibrosis. Maternal prognosis is better with non-cirrhotic portal hypertension compared to cirrhosis of liver.

The causes of death are generally hematemesis, hepatic coma or postpartum hemorrhage. The mother is also at risk of developing severe anemia, ascites, splenic artery aneurysm rupture, spontaneous bacterial peritonitis. Perinatal mortality ranges between 11-18%. Variceal bleed is the most morbid complication and the gold standard to assess the risk of bleeding is Endoscopy. Current American Association for the Study of Liver Disease (AASLD) recommendations include screening endoscopy in 2nd trimester as there is maximum rise in portal pressure.

Prinicipal risk in patients on Non-selective betablockers is fetal bradycardia, fetal growth restriction for which serial monitoring has to be done and prevented. Current literature (Baveno V consensus workshop) recommends EVL (Endoscopic variceal ligation) for acute variceal bleed, sclerotherapy if banding is difficult.

Upper GI endoscopy in our patient revealed grade III esophageal varices as per the Kodsi classification, characterized by medium caliber varices extending into the middle third of the esophagus.

Figure 2: Kodsi grading of esophageal varices.

Grade	Endoscopic findings
0	Absence of esophageal varices
I	Microvessels that sketch varicose strings located in the esophagogastric transition or in the distal esophagus
II	One or two fine-caliber varices (smaller than 3 mm diameter) located in the distal esophagus
III	Medium caliber varices (between 3 or 6 mm diameter) or more than varices up to 3 mm that may reach up to a third medium third of the esophagus.
IV	Thick caliber varices, larger than 6 mm diameter, in any part of the esophagus.

Figure 3: Endoscopic images of esophageal varices - Grades I to IV as per Kodsi classification.

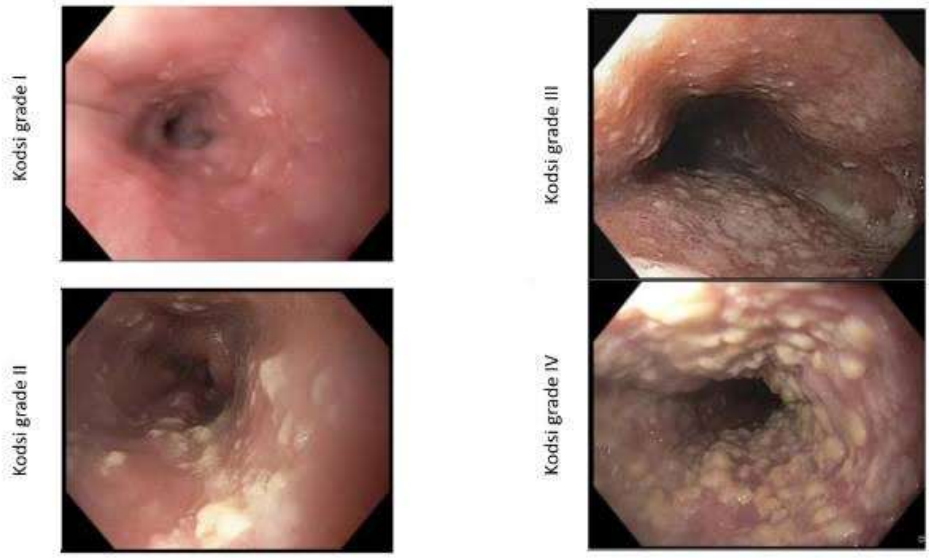
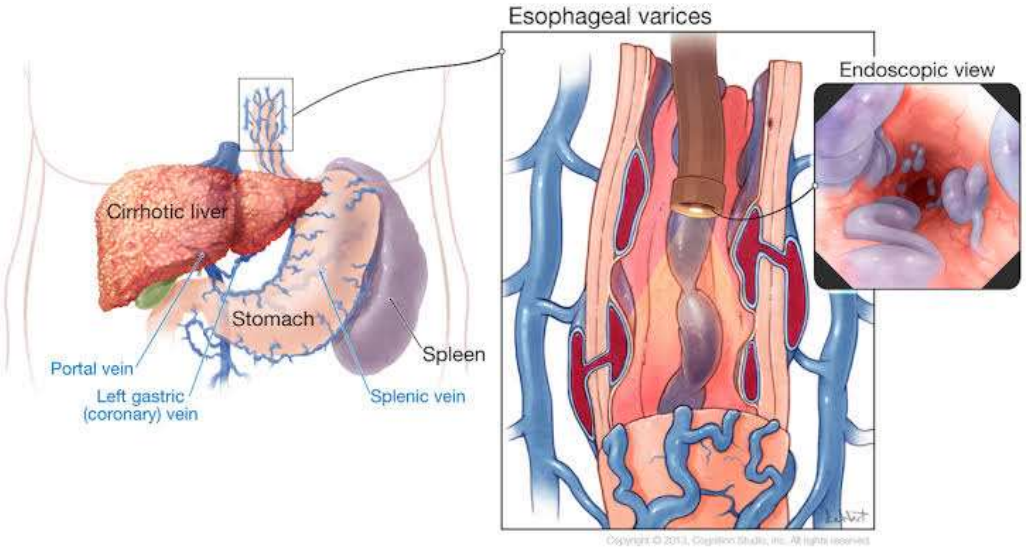


Figure 4: Schematic diagram showing the anatomical basis of portal hypertension and esophageal varices due to cirrhotic liver.



Conclusion

This case highlights the need for heightened vigilance and early endoscopic evaluation in pregnant women presenting with esophageal varices. Pre-conceptional counseling, regular follow-up and monitoring are essential. Multidisciplinary care involving gastroenterologists, obstetricians, hepatologists, hematologists, anesthesiologists at a tertiary care level is paramount to ensure optimal outcomes.

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