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Case Report

A rare presentation of non-cirrhotic portal fibrosis in pregnancy: a case report

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ABSTRACT

Non cirrhotic portal fibrosis (NCPF) in pregnancy is a rare occurrence with an in hospital incidence of 0.009%. Management in pregnancy is challenging due to lack of standard protocol and paucity of extensive research in current literature. NCPF if untreated can lead to catastrophic complications and lead to significant maternal morbidity and mortality. We present a case report of a 23-year-old G2A1 female presenting for the first time in pregnancy at 34 weeks' gestation as a case of NCPF with prelabour rupture of membranes. She was managed with multidisciplinary approach and underwent normal vaginal delivery with satisfactory feto-maternal outcome. NCPF in pregnancy is uncommon and a thorough knowledge about the condition and systematic management at a tertiary care center with a multidisciplinary team is essential. Its prognosis in pregnancy is usually good if managed promptly and systematically.

Keywords: Non cirrhotic portal fibrosis, Pregnancy, Portal hypertension

INTRODUCTION

Portal hypertension is defined as a pressure gradient of 3.5 mm Hg between the inferior vena cava and portal vein. It can have either cirrhotic or non-cirrhotic etiology. There are multiple causes of non-cirrhotic portal hypertension. However, non-cirrhotic portal fibrosis (NCPF) and extra hepatic portal vein obstruction (EHPVO) are the two most common causes of non-cirrhotic portal hypertension. Non cirrhotic portal fibrosis is a condition characterized by periportal fibrosis and involvement of small and medium sized branches of portal vein.[1] It is associated with portal hypertension, massive splenomegaly and variceal bleeding with preservation of hepatic function. Prevalence of NCPF in general population is 3-5% and 15-20% in world and India respectively. Its occurrence in pregnancy is rare with an in hospital incidence of 0.009%. Its management in pregnancy is challenging because of paucity of extensive research in this area and lack of standard practice guidelines in literature.

CASE REPORT

This is a case report of a 23-year-old G2A1 female, who presented at 16 weeks' gestation in antenatal clinic with complaint of pain abdomen and jaundice. Her general physical examination was unremarkable. Abdominal examination revealed splenomegaly with lower edge of spleen felt 8 cm below left costal margin. Liver was not palpable. Gravid uterus corresponding to 16 weeks' gestation was noted. Her baseline investigations were as follows: haemoglobin 13 gm/dl, total leucocyte count 13000, platelet count 22,000/microlitre, total bilirubin 0.42 mg/dl, direct bilirubin 0.10 mg/dl, aspartate aminotransferase (AST) 231.10 U/l, alanine aminotransferase (ALT) 159.00 U/l, alkaline phosphatase (ALP) 258 U/l, prothrombin time (PT) 13.4, international normalized ratio (INR) 1.1, blood group A positive, human immunodeficiency virus (HIV), veneral disease research laboratory test (VDRL), hepatitis B virus (HbsAg), and hepatitis C virus (HCV-Ab) were negative. Serology for viral hepatitis A, B, C and E was non-reactive. Urgent

review in gastroenterology department was done. Direct Coombs test, antinuclear antibody (ANA) and liver line assay was negative. Ultrasound whole abdomen and revealed no distortion of liver architecture, dilated portal vein (22 mm), dilatation of portal collaterals, ascites with enlarged spleen (15 cm). Upper gastrointestinal (GI) endoscopy revealed grade 2 varices. Her bone marrow aspirate findings were unremarkable. There were no clinical features of Wilsons disease. Liver biopsy could not be done due to thrombocytopenia.

She was diagnosed as a case of NCPF since five years. She presented with pain abdomen 5 years back to gastroenterology department of our facility. Her ultrasound whole abdomen and contrast enhanced computed tomography (CT) revealed no distortion of liver architecture, dilated portal vein (22 mm), dilatation of portal collaterals, ascites with enlarged spleen (15 cm) with pancytopenia. Serology for viral hepatitis A, B, C and E was non-reactive. DCT, ANA and liver line assay was negative. Upper GI endoscopy revealed grade 1 varices. Her bone marrow aspirate findings were unremarkable. Serum ceruloplasmin was not done due to patient's financial constraints. However, there were no clinical features of Wilsons disease. Liver biopsy was not done due to severe thrombocytopenia. She was following up regularly at gastroenterology department.

She had a spontaneous first trimester abortion 5 years back which was not followed by D&C. Her current conception was spontaneous and antenatal period was uneventful, with no episodes of fever, jaundice, epistaxis and haematochezia. There was no history of taking drugs like beta blockers or nitrates in the past. Her pregnancy was unbooked and unsupervised. After conceiving, she presented for the first time in antenatal clinic at 34 weeks with prelabour rupture of membranes. General physical examination was unremarkable. Abdominal examination revealed splenomegaly with lower edge of spleen felt 8 cm below left costal margin. Liver was not palpable. Gravid uterus corresponding to 34 weeks' gestation was noted. She was admitted and baseline investigations were as follows: haemoglobin 13 gm/dl, total leucocyte count 13000, platelet count 000/microlitre, total bilirubin 0.42 mg/dl, direct bilirubin 0.10 mg/dl, AST 21.10 U/l, ALT 19.00 U/l, ALP 158 U/l, PT 13.4, INR 1.1, blood group A positive, HIV, veneral disease research laboratory test (VDRL), hepatitis B virus (HbsAg), and hepatitis C virus (HCV-Ab) were negative. The obstetric ultrasound gave an estimated fetal weight of 2100 g with a biophysical profile score of 8/8. A multidisciplinary discussion was held including gastroenterology, haematology, neonatology, anaesthesiology and obstetrics. It was decided that as there was no history of variceal bleeding in the past and with presence of low-grade varices, it would be optimal to proceed for an induction of labour without therapeutic UGIE. She was tranfused single donor platelets and underwent induction of labour with dianoprostone gel. After two doses of dianoprostone gel, she progressed to active labour after 18 hours. She delivered a 2215 grams male baby vaginally without any instrumental aid. Active management of third stage of labour was done and estimated blood loss was 400 ml. Postpartum period was uneventful. She was given routine antibiotics in post-partum period. Contraceptive counselling was done. She opted for barrier contraception. Baby had an APGAR of 8, 8, 8 and had normal course after birth.

DISCUSSION

Non cirrhotic portal fibrosis in pregnancy is a rare occurrence. While cirrhosis is the main cause of portal hypertension in the west, NCPF and EHPVO are more common causes of portal hypertension in India. Literature supporting NCPF in pregnancy is limited to 3 case reports and 4 observational studies (Table 1).[2-8]

An observational study conducted by Kochar et al in 55 women with non-cirrhotic portal hypertension evaluated fertility rate, fetal losses and variceal bleeding and found no significant difference in these outcomes between women diagnosed with portal hypertension before or after pregnancy and controls.[2] Andrade et al reported favorable fetal outcomes but significant portal hypertension related complications in their retrospective observational study including 16 women with idiopathic non cirrhotic portal hypertension (INCPH).[3] A retrospective observational study, recruiting 27 women with INCPH revealed that upper GI bleeding was more common among women with EHPVO as compared to women with NCPF (43.9% versus 25.9%). However, perinatal adverse outcomes were noted to be more common among women with NCPF as compared to EHPVO.[4] Conflictingly, comparison between obstetric and feto-maternal complications between NCPF and EHPVO revealed no significant difference in another observational study conducted in South India. Out of 108 pregnancies, 74.1 % women had NCPF.[5]

Thus, there is a lot of heterogeneity in existing literature regarding feto-maternal prognosis in women with NCPF. Management is equally challenging and lacks good quality evidence. NCPF is usually idiopathic but it may be associated with conditions like hematological malignancies, immunological disorders, chronic infections, thrombophilias and genetic disorders.[9] These conditions were ruled out in our case and NCPF seemed to be idiopathic. Histopathologic characteristics include hepatoportal sclerosis and nodular regenerative hyperplasia.[9]

Fertility of women in NCPF is maintained and conception is usually spontaneous. Women with NCPF usually present with variceal bleed, ascites and splenomegaly. Course of disease during antenatal period depends on various factors. Previous history of variceal bleed, present jaundice, hypersplenism and presence of large varices on endoscopy are factors that are predictive of variceal bleed during pregnancy. Women with previous history of variceal bleed have 78% chance of repeat bleeding during pregnancy.[10] There is 29.4% chance of abortion and 33.3%

chance of perinatal death in women experiencing variceal bleed during pregnancy.[11] Hypervolemic state of pregnancy further aggravates portal hypertension and increases chances of variceal bleed during pregnancy. Variceal bleeding can be catastrophic during pregnancy and it is a predictor of poor feto-maternal outcome. In case of acute bleed in pregnancy, immediate rescucitation followed by upper GI endoscopy is the dictum. Endoscopic variceal ligation (EVL) is the mainstay of treatment and is effective in 80-90% women in controlling acute variceal bleed. Endoscopic sclerotherapy is also reported in few cases, however EVL is preferred choice to avoid risks of sclerosants in pregnancy. Medical therapy with vasopressors for acute variceal bleed has a role, however their safety in pregnancy is questionable. Beta blockers may be used to reduce the portal venous pressures and for prophylaxis of variceal bleed.

During intrapartum period, vaginal delivery is usually well tolerated in women with NCPF and caesarean section is reserved for obstetric indications. However, risk of variceal bleed may increase if the patient strains in labour. Therefore, epidural analgesia during labour preferable with aid of force or ventouse extraction. Another novel approach, reported in 2017 describes combined caesarean section with splenectomy for patients with transfusion dependant pancytopenia secondary to hypersplenism in a woman with NCPF.[6] Further studies are needed to investigate this approach further.

Due to preserved liver function, NCPF is generally associated with better prognosis and fertility as compared to women with cirrhotic disease. However, there are many unanswered questions that are yet to be answered through adequately powered studies.

CONCLUSION

NCPF is a rare condition in pregnancy but can lead to catastrophic complications if it remains unidentified and untreated. A thorough knowledge about the condition and systematic management at a tertiary care center with a multidisciplinary team is essential. Its prognosis in pregnancy is usually good if managed promptly and systematically.

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