CASE REPORT

Successful Pregnancy Outcome with Extra Hepatic Portal Venous Obstruction: Three Case Series

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About the Author

Nilesh Mhaske is a postgraduate student in LTMGH and has been taking keen interest in research related to Obstetrics and Gynecology. His future interests include community obstetrics and ensuring implementation of standard practices in low-resource settings.

Introduction

Pregnancy associated with portal hypertension is a high-risk condition as both pregnancy and portal hypertension contribute to worsening of hemodynamic status. The adaptive changes worsen the portal hypertension and can result in potentially life-threatening variceal bleeding and associated

complications. The liver function is much better preserved in non-cirrhotic portal hypertension than in those associated with cirrhosis. The exact incidence of non-cirrhotic portal hypertension in pregnancy is not known [1]. The management of such patients requires multidisciplinary approach, preferably in a tertiary care institution. Three such successfully managed patients are discussed here.

Cases

Case 1

A 32-year-old primigravida, married since 12 years, presented at 28-week gestation with loose motions for 2 days and swelling of feet since 2 weeks. Patient had undergone treatment for infertility, leading to conception. Pallor was present. Facial, vulval and bilateral pedal edema was present. There were no signs of hepatocellular involvement.

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Fig. 1 Ultrasonography showing splenomegaly

Obstetric examination was within normal limit. For pancytopenia with severe hypo-proteinemia, 1 unit of blood and 2 units of 20 % human albumin were transfused. Other liver and kidney parameters were within normal limit. Urine examination showed nephrotic range proteinuria and gross hematuria. DCT/ICT was negative. Ultrasonography showed splenomegaly (Fig. 1) with multiple dilated collaterals within the splenic hilum; chronic thrombosis of portal and splenic vein; altered liver echo texture and minimal ascites suggestive of EHPVO. Specialist gastro-medicine and nephrology opinion was taken for further management. Surveillance endoscopy showed no esophageal or gastric varices. ANA, LA, aCLA were negative, and ASO titer was normal. Compliment C3, C4 levels were low, and hence, patient was started on Tab. deflazacort 30 mg once a day and followed up regularly in antenatal outpatient department and a post-delivery renal biopsy was planned. Patient developed pre-eclampsia at 30 weeks of gestation requiring anti-hypertensives. Patient went into spontaneous labor at gestational age of 34 weeks. Ten units platelet transfused peri-delivery and a female child of weight 1560 g delivered vaginally. Hematological, hepatic and renal parameters dramatically improved around 2 weeks post-delivery, and hence, renal biopsy was deferred. Patient was kept under observation, also because baby was in NICU. Patient was discharged on day 26 of delivery, with complete resolution of symptoms.

Case 2

A 28-year-old primigravida married since 1 year presented at 30 weeks of gestation with deranged sugars. EHPVO had been diagnosed at the age of 9 years and was on oral propranolol 40 mg. She underwent upper intestinal

Fig. 2 Colour Doppler showing narrowed portal vein with peri-portal collaterals

endoscopy multiple times, requiring sclerotherapy thrice. At 18 weeks of gestation, two large esophageal varices necessitated variceal ligation.

On general examination pallor was present. There was no icterus, edema or signs of liver cell failure. On per abdominal examination, massive splenomegaly was present. Obstetric examination was within normal limit. Investigations revealed pancytopenia and deranged blood sugar levels, while liver and kidney function tests were within normal limit. Ultrasonography revealed altered liver echostructure with central intra-hepatic biliary radicle dilatation, and portal vein was replaced by multiple dilated collaterals in the peri-portal area (Fig. 2). Gastro-medicine specialist reviewed and advised no specific intervention. Subcutaneous insulin was required to control blood sugars. Induction of labor was done at 39 weeks, and due to non-progress of labor, LSCS was done, with peri-operative platelets and FFP transfusion, and delivered a healthy male child of 3.1 kg. Post-LSCS, she required 3 units of blood transfusion. Patient developed subcutaneous wound hematoma which resolved spontaneously and severe ascites requiring 1 unit of 20 % human albumin transfusion.

Case 3

A 31-year-old primigravida, married since 1 year, was a diagnosed case of EHPVO. Patient had one episode of hematemesis 7 years back, requiring variceal ligation and was on tab. propranolol 40 mg since then. Endoscopy at 20-week gestation was suggestive of esophageal varices with portal hypertensive gastropathy (Figs. 3, 4) and was advised variceal ligation. However, patient defaulted and

Figs. 3 and 4 Endoscopic picture showing esophageal varices and portal hypertensive gastropathy

Fig. 5 Colour Doppler showing prominent hepatic artery

presented at 34 weeks of gestation. On general examination, pallor was present. There was no icterus, edema feet or signs of liver cell failure. On per abdominal examination, massive splenomegaly was present and obstetric examination was within normal limits. Blood investigations revealed pancytopenia, with normal liver and kidney function tests. There was no associated proteinuria or hematuria. Protein C, protein S, LA, aCLA, Factor V and Factor VIII were negative. Ultrasonography revealed caudate lobe hypertrophy, portal vein obstruction and severe splenomegaly with mild ascites (Fig. 5). With peri-operative platelet transfusion, elective LSCS was done at 39 weeks of gestation for breech presentation and delivered a male child of 3 kg. Post-operative period was uneventful.

Discussion

EHPVO is a rare condition, especially in the geographical location from where this study is being reported. The etiological factors associated include home delivery and umbilical sepsis [2]. EHPVO in pregnancy is rare, and very few studies pertaining to it have been reported [3].

The mean maternal age in our study was 30 years, whereas the mean age in a large case series was 24 years. The disease was diagnosed in this pregnancy in only one of the three. In a large case series, 55 % of the patients were diagnosed during the current pregnancy. Though none of our patients had variceal bleeding during pregnancy, a 45-patient series reported bleeding only in three patients [4]. The occurrence of the same has been associated with a higher incidence of abortion and perinatal death [5].

Two of our three patients went to term gestation. A term birth is reported in 58 % of such pregnancies [4]. Two of our patients required Cesarean section, both for obstetric indications. Cesarean section was used in 53 % of deliveries in a large case series [4]. In our study, the mean birth weight was 2553 g. In a large study, it was around 2668 g [5]. All three of our patients required peri-delivery platelet transfusion. In a large study, 50 % of the patients required platelet transfusion [6].

Overall, the outcome was favorable in all three patients. An overall favorable outcome has been reported in all case studies, and no maternal deaths have been reported [3–6].

Conclusion

Pregnancy with portal hypertension is not contraindicated as was once believed. With better diagnostic and treatment modalities, maternal and fetal outcomes are constantly

improving. Physicians and obstetricians need to coordinate well regarding individualization of treatment and be observant regarding specific risks, which include hemorrhage (mainly due to variceal bleeding, aneurysmal rupture and PPH) or hepatic failure. A multidisciplinary team approach in a tertiary care set-up, which resulted in three successfully managed patients, is reported here. Further prospective studies are needed in this regard.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Standards All authors declare that all applicable ethical standards have been complied with.

Human and Animal Rights This study did not involve any use of animals.

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