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CASE REPORT

A Rare Case of Portal Vein Cavernoma with Pregnancy

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ABSTRACT

Portal vein cavernoma is a rare disease resulting from extra-hepatic portal vein thrombosis and development from collateral venous circulation. It is a rare condition affecting both adults and children with equal gender distribution. Both local and systemic factors may contribute to development of portal vein thrombosis. Once there is portal vein thrombosis, cavernous transformation occurs in 5 weeks to 12 months of thrombotic event. We report a case of primigravida with portal vein cavernoma who presented to us in last trimester. Cesarean section was done with primary indication with premature rupture of membrane with poor bishop score. Cesarean section was given priority considering associated digestive varices. Both maternal and fetal outcome was good.

Keywords: Portal vein cavernoma, Portal vein thrombosis, Pregnancy.

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INTRODUCTION

Portal vein cavernoma is a rare disease resulting from extrahepatic portal vein thrombosis and development of portal venous channels. The management of pregnancy and delivery in woman with portal vein cavernoma has rarely been described. We are reporting this case for its rarity.

CASE REPORT

A 24 years old, primigravida reported in antenatal OPD with history of amenorrhea since 9 months. She had no other complaints. She was unbooked patient. There was no history suggestive of raised blood pressure, edema feet, bleeding per vaginum. Patient gives history of acute pain in abdomen around 28 weeks of gestation for which she did not consulted to any doctor. She had no history of hematemesis. She had no complaint of pain in epigastric region. Her history was uneventful. There was no history of use of oral contraceptive pills. On general and systemic examination, she was afebrile. Her vitals were stable with pulse of 88/min regular and blood pressure of 120/80 mm Hg. Respiratory and cardiovascular system was normal. There were no signs suggestive of bleeding tendencies. There was no hepatosplenomegaly. Obstetric examination revealed singleton pregnancy corresponding to the weeks of gestation with vertex presentation and clinically mild oligohydramnios. Fetal heart rate was normal. Patient was admitted in view of 40 weeks of gestation with oligo-hydramnios. On investigations ultrasonography revealed portal vein cavernoma (Figs 1 and 2) with single live intrauterine pregnancy corresponding to week of gestation with mild oligohydramnios. There was no splenomegaly, no mesenteric vein thrombosis. Liver size and echotexture was normal. In blood investigations, her total and differential leukocyte count was normal (see Figs 1 and 2). Platelet count was 3 lakh/cm. Her coagulation profile was normal. Physician's opinion was taken. Patient was advised for endoscopy to rule out digestive varices. She was advised investigations for antiphospholipid antibodies, serum homocystein levels, protein C, S, antithrombin III levels and test for Factor V Laiden mutation. Anticoagulation therapy was not started as patient presented to us at term. Meanwhile patient had premature rupture of membrame. Patient was taken for cesarean section in view of poor bishop score and suspected digestive varices. Blood and blood products were kept ready. The intraoperative and postoperative period was uneventful. She delivered a healthy

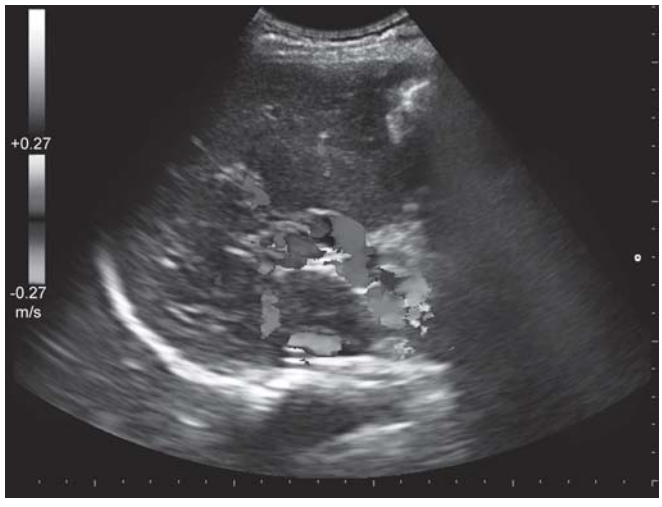

Fig. 1: Cavernoma USG

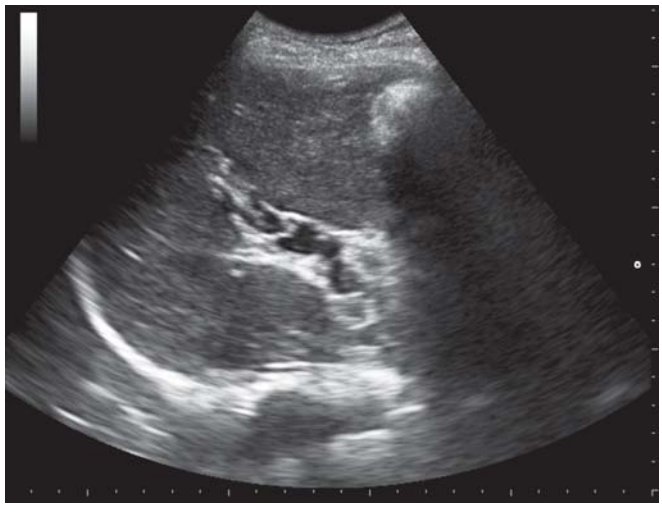

Fig. 2: Portal vein cavernoma USG

baby weighing 2.6 kg. Before discharge from hospital, her gastrointestinal endoscopy was done which revealed no digestive varices. Mother and baby were discharged on 9th day in a good condition. We could not rule out the cause for portal vein thrombosis and subsequent cavernoma formation as patient was not willing to get herself investigated for the same at that time. She was willing for investigations in follow-up visit. But, patient did not come for follow-up thereafter.

DISCUSSION

Portal vein cavernoma is a rare disease resulting from extra-hepatic portal vein thrombosis and development from collateral venous circulation. GW Balford and TG Stewart in 1869 first reported portal vein thrombosis (PVT) in a patient who presented with ascites, splenomegaly and varices.[1] It is a rare condition affecting both adults and children with equal gender distribution. Though overall incidence ranges from 0.05 to 0.5% of autopsy studies in US, high incidence is noted in a patient with cirrhosis of liver.[1] Once, there is PVT, cavernous transformation (portal vein cavernoma—PVC) occurs in 5 weeks to 12 months of thrombotic event.[1] The overall consequence of PVT is related to thrombus extension. Anatomically, it is classified into four categories according to where the thrombus extends. Both local and systemic factors may contribute to development of PVT. Common local etiological factors being cirrhosis, portal hypertension, prothrombotic tendencies like myeloproliferative disorders, antiphospholipid antibodies, anticardiolipin antibodies, thrombophilias (protein C, S and antithrombin III deficiency), Factor V Leiden mutation, hyperhomocysteinemia, paroxysmal nocturnal hemoglobinuria, local or distant malignancies, local or systemic sepsis (Bacteroid fragilis), pancreatitis, schistosomiasis, postsurgical (splenectomy), portal vein thrombosis by nodes (tuberculosis, lymphoma), drugs (oral contraceptive pills), pregnancy and postpartum period.[2] As pregnancy is prothrombotic condition, it predisposes pregnant women for the development of PVT. The patient's presentation can be acute or chronic. Acute PVT presentation there is abdominal pain, nausea, fever and mostly symptoms are because of mesenteric venous thrombosis and associated bowel ischemia. Chronic PVT might present with esophageal or gastric varices.[2] A range of imaging modalities may be used in diagnosis of PVT and PVC, like ultrasonography, Doppler ultrasonography, computerized tomography and MRI.[1,2,4]

Treatment is surgical and medical. Surgical treatment includes shunts, band ligation of esophageal varices. Medical treatment includes use of B-blockers to decrease the flow in portal circulation.[2] Anticoagulation therapy should be given as studies have suggested that it results in recanalization in more than 80% cases.[2] Very few cases of PVC with pregnancy have been reported. There are two case reports from France in 2009.[3] First patient presented with PVC with pregnancy with large jejunal varices and high anticardiolipin antibody. She was delivered by cesarean section. The second patient had protein C deficiency and thrombocytopenia delivered vaginally without any complications. Both were treated with B-blockers and low molecular weight heparin.[3] The another case reported was from USA in 2006. This patient presented as primigravida with 28 weeks of gestation with cavernous transformation of portal vein. Esophageal and gastric varices were ruled out by endoscopy. She was delivered by cesarean section.[4] Other retrospective multicentric analyses between 1995 to 2008 reported 10 cases.[5] In pregnancy with PVC patients, a fetal outcome is favorable. Maternal outcome is also good on anticoagulation.[5] However, anticoagulant therapy should be stopped during term for safe delivery and cesarean section as it is anticipated with increased bleeding. Elective cesarean seemed necessary in cases with digestive varices. Vaginal delivery with passive second stage, seems to be relatively safe and less morbid in women without digestive varices.[3]

Our patient consulted late in pregnancy. Hence, anticoagulation was not started. Patient gives history of acute pain in abdomen at 28 weeks of gestation. That might be the acute episode of portal vein thrombosis which subsequently was transformed into cavernoma. She was delivered by cesarean section for obstetric indication and possibility of digestive varices in view of portal vein cavernoma. Both maternal as well as neonatal outcome was good in our case also.

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