

Pregnancy with Maternal Caroli's Disease - Case Report

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Introduction: - Caroli's disease is a rare congenital disorder characterized by multifocal, cystic dilatation of large intrahepatic bile duct [1, 2]. The disease affects about 1 in 1,000, 000 people. Most cases are transmitted in autosomal recessive fashion. Dilatation of bile ducts predispose to stagnation of bile, leading to the formation of biliary sludge and intraductal lithiasis. Bacterial cholangitis occurs frequently. Patient can present with portal hypertension secondary to fibrosis of liver. Coagulopathy from Vitamin K malabsorption may occur in cholestasis patient [3]. Because of rarity of disease, data on pregnant women with Caroli's disease is limited. To our knowledge, this case is the fourth report about Caroli's disease in pregnancy [4, 5, 6].

Case presentation: - The 30 years old woman, gravida 2, para 1 with no live child, presented to us for her antenatal care. In her previous pregnancy she conceived by ovulation induction medicine and had

intrauterine death at 25 weeks with placental abruption. She received blood transfusion and fresh frozen plasma. She had secondary postpartum hemorrhage on postpartum day 3 for which uterine artery embolization was done. At that time she was diagnosed with Caroli's disease. Liver function test and coagulation parameters were deranged. MRCP and MRI were suggestive of Caroli's disease, dilated intra and extra hepatic biliary with intraductal lithiasis.

She presented to us in her second pregnancy at 8 weeks. In collaboration with a hepatologist, she was managed on ursodeoxycholic acid along with routine antenatal care. Every month her liver function test and coagulation profile were checked and vitamin K injection was given. At 30 weeks she came to us with complains of pain in upper abdomen, ribs, back and fever. The

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Running title

Maternal Caroli's disease and pregnancy.

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ultrasound findings were suggestive of primary sclerosing cholangitis. She was managed conservatively on intravenous fluids, antibiotics (3rd generation cephalosporin) and vitamin K injection. On the same day she also developed preterm uterine contraction and hence tocolytic agents along with antenatal corticosteroid for fetal lung maturity were given. She improved symptomatically and was discharged then.

At 32 weeks she reported to us with bleeding per vaginum. The baby was in breech presentation.

Investigations: - On presentation the cardiotocography (CTG) was normal (baseline Fetal heart rate 140 beats per minute). Her haemoglobin was 12.4gm/dl. Her coagulation profile was deranged with INR-1.69, PT 17.5/13.2 (test/control) and APTT (48.9/37.9) respectively. The results of her liver function test were bilirubin total 1.9mg/dl, direct 1.4 mg/dl, SGOT-62U/L, SGPT 76U/L, AIKPO4-446 U/L. The ultrasound findings were single live fetus, 31 weeks and one day gestational age, estimated fetal weight 1516 gms +/- 15%, with breech presentation, , placenta being at upper segment of anterior wall, no retroplacental collection, normal umbilical cord, with normal obstetric doppler , and cervix length 2.55cm.

Treatment: - Her treatment on presentation included intravenous rehydration and intramuscular injection vitamin K. As the patient had history of bleeding with breech presentation emergency caesarean section was done. Four units of fresh frozen plasma were transfused perioperatively. Placenta was sent for histopathological examination. Uterus, bilateral tubes and ovaries were normal looking with no abnormal vascularisation. A live baby girl weighing 1510 gms was delivered. As the baby was preterm, she was shifted to NICU.

Outcome and Follow-up: - Her recovery was uneventful and she was discharged home on the 4th postoperative day. Her treatment at discharge included antibiotics, painkiller along with ursodeoxycholic acid.

The baby was in NICU for 30 days. She required continuous positive airway pressure for 3 days. At the time of discharge her weight was 2050 gms and she was accepting breast feeding.

Discussion: - Caroli's disease usually presents during childhood and early adulthood. Patients usually present with right upper abdominal pain, fever or jaundice due to bacterial cholangitis or hepatolithiasis. Treatment includes supportive care with antibiotics for cholangitis and ursodeoxycholic acid for hepatolithiasis [3, 4].The course of pregnancies complicated by Caroli's disease are variable and can include life- threatening conditions like acute ascending cholangitis, disseminated intravascular coagulation and septic shock as reported by Adair et al [5]. In his case study fetal distress necessitated delivery by caesarean section, followed by mother's postoperative course that required prolonged critical care and multidisciplinary care. Kejariwal and Sarkar [4] reported Caroli's disease , simple type with diffuse bile duct ectasia with uneventful pregnancy. Tsunoda M et al [6] reported a case of Caroli's disease associated with chronic renal failure due to polycystic kidney disease, with an uneventful pregnancy course. The present case demonstrates that maternal complication of Caroli's disease can affect the course of pregnancy. Her abdominal pain due to cholangitis led to preterm uterine contraction, which was managed with tocolytic agents, antibiotics and antenatal corticosteroid injection.

During caesarean section, placenta was removed by controlled cord traction; it was normal looking with no clot underneath it. There was no postpartum haemorrhage. The likely explanation for this antepartum haemorrhage is deranged coagulation profile secondary to Caroli's disease. Histopathology of the placenta revealed normal placenta with no evidence of thrombosis, ischemic necrosis, choriovillitis or abnormal placentation.

Following uterine artery embolization, uterine hypovascularisation could affect placentation, fetoplacental exchanges and subsequent fetal growth, further leading to vascular complications like preeclampsia, intrauterine growth restriction etc [7, 8, 9]. All these complications were not found in our case study. Her normal conception after uterine artery embolization reinforces and encourages possibility of future pregnancy after uterine artery embolization.

In conclusion though Caroli's disease is a rare disorder and can be fatal for both mother and child but with

regular antenatal care and proper management, good outcomes can be expected. The present case thus emphasizes the importance of multidisciplinary care, timely intervention and close follow-up.

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