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Primary sclerosing cholangitis revealed during pregnancy: case report

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Abstract

Primary sclerosing cholangitis (PSC) is a cholangiopathy characterized by chronic fibroinflammatory damage of the biliary tree and is frequently associated with inflammatory bowel disease (IBD). Its association with pregnancy is exceptional and presents unique diagnostic and therapeutic challenges. We report a case of PSC revealed during pregnancy. It's about a 30-year-old woman, gravida 1, para 0, with no significant medical history, presented at 16 weeks of pregnancy with intense pruritus and persistent fatigue. Laboratory findings revealed cholestasis with elevated alkaline phosphatase, gamma-glutamyl transferase, and transaminases. An abdominal ultrasound ruled out cholelithiasis, while MRCP demonstrated multifocal bile duct strictures, suggestive of PSC with a chronic liver disease. The patient was managed with Ursodeoxycholic acid and close multidisciplinary monitoring. Delivery occurred at term without major complications, and postpartum follow-up was established to monitor disease progression.

This case highlights the importance of considering PSC in atypical gestational cholestasis and underscores the need for a multidisciplinary approach to optimize maternal-fetal management.

Introduction:

Although liver disease is rare during pregnancy, it can pose a significant risk to both the mother and fetus. Pregnancy-related liver disorders include preeclampsia, HELLP syndrome (hemolysis, elevated liver enzymes, low platelets), obstetric cholestasis, hyperemesis gravidarum, and acute fatty liver disease. Diagnosing liver disease in pregnancy can be challenging due to overlapping symptoms. One rare cause of liver disease during pregnancy is primary sclerosing cholangitis, a condition characterized by chronic inflammation, destruction, and fibrosis of intrahepatic and extrahepatic bile ducts. Due to the bile ducts' limited ability to regenerate, the disease often progresses to cirrhosis, portal hypertension, and liver failure. It is more

common in men than women, and typically develops between the ages of 30 and 60, though it can begin in childhood. Symptoms such as itching, fatigue, jaundice, and weight loss indicate disease progression, although early histological changes may be asymptomatic. Diagnosis is made using magnetic resonance cholangiopancreatography (MRCP), sometimes by liver biopsy in PSC of small ducts. While ursodeoxycholic acid (UDCA) has not been proven to alter the disease's course, it appears to improve symptoms and laboratory findings. In end-stage primary sclerosing cholangitis, liver transplantation remains the only life-extending treatment.

We present the case of a woman who reveled primary sclerosing cholangitis during pregnancy.

Case report:

A 30-year-old primiparous woman, consulted during the second trimester of her pregnancy due to generalized pruritus and was both isolated and disabling.

The initial clinical examination revealed a blood pressure of 107/70 mmHg, a negative urine test, and no temperature. Scratching lesions were observed on her abdomen, upper, and lower limbs, with no obvious dermatological cause.

Laboratory tests showed a normal level of hemoglobin, platelets at 170,000/mm³ (150,000–400,000), white blood cells at 7040/mm³ (4,000–10,000), ALAT at 420 UI/L (0–35), ASAT at 147 UI/L (0–35), total bilirubin at 72 mg/L (3–12), and direct bilirubin at 50 mg/L (0–2). Alkaline phosphatases were measured at 192 UI/L (30–120), gamma-glutamyl transferase at 76 UI/L (0–38), and the prothrombin level was 100%, with normal albumin levels. Bile acid testing returned a value of 48.4 µmol/L (<8).

Hepatobiliary ultrasound revealed chronic liver disease without bile duct dilation or lithiasis.

Urine cytobacteriological examination and serologies for chronic hepatitis B and C were negative. The immunological assessment, including tests for anti-mitochondrial antibodies, anti-smooth muscle AC, anti-nuclear antibodies, anti-LKM1, and anti-ALS, was also negative.

Further imaging with biliary MRI showed a chronic liver condition with multi- focal strictures and dilatations of the intrahepatic bile ducts (Figure 1), suggesting primary sclerosing cholangitis with signs of portal hypertension.

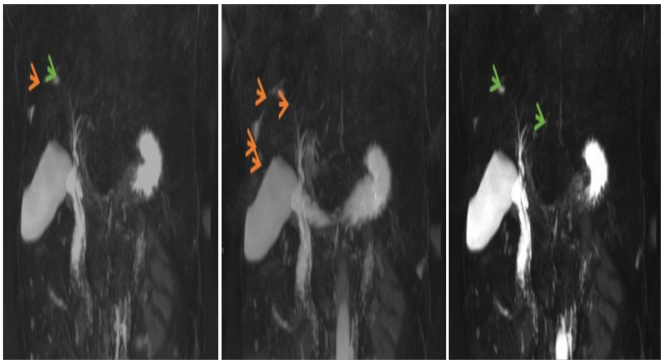

Figure 1: Coronal Bili MRI showing areas of focal narrowing of the intrahepatic bile ducts (orange arrows) associated with areas of focal dilatation (green arrows).

Secondary causes of sclerosing cholangitis were ruled out, confirming the diagnosis of primary sclerosing cholangitis at the chronic liver disease stage. A screening gastroscopy detected small esophageal varices.

Regarding obstetric findings, fetal movements were well perceived, there were no fetal heart rate

abnormalities, and obstetric ultrasound showed a single progressive pregnancy with normal amniotic fluid volume.

The patient was treated with ursodeoxycholic acid (UDCA) at an initial dose of 15 mg/kg/day (250mg/day), with gradual dose escalation up to 750 mg/day. Pregnancy follow-up was conducted in collaboration with a gynecologist, focusing on clinical (pruritus intensity), biological (liver function tests every 2–3 weeks), and fetal monitoring (fetal movements, fetal heart rate, and obstetric ultrasound).

The pregnancy progressed without complications, with a significant improvement in pruritus and liver function tests. From the 37th week, monitoring was intensified with bi-weekly fetal assessments. Labor was induced at 39 weeks of gestation, but due to induction failure, the patient. ultimately delivered via cesarean section, giving birth to a healthy male newborn weighing 3,800 g.

Additionally, a postpartum colonoscopy ruled out any association with inflammatory bowel disease.

Discussion:

Primary sclerosing cholangitis may represent a severe disruption of maternal-fetal bile salt metabolism, like obstetric cholestasis. The proposed mechanism of obstetric cholestasis is linked to hormonal influences, particularly increased estrogen levels, which impact the bile salt export pump and the basolateral membrane of hepatocytes. This disruption impairs the transfer of bile acids from the mother to the fetus across the placenta, potentially leading to toxic bile acid accumulation in the fetus. Elevated bile acid levels may also affect myometrial contractility and induce vasoconstriction of chorionic veins in the placenta, which could contribute to preterm labor and fetal distress. Mutations in hepatocyte transporter genes may act as disease modifiers in primary sclerosing cholangitis, and the interaction of these mechanisms could potentially trigger severe exacerbations of primary sclerosing cholestasis. The management of idiopathic cholestasis of pregnancy involves two main approaches: symptomatic treatment with ursodeoxycholic acid (UDCA) and timely fetal delivery. UDCA alleviates maternal pruritus and improves cholestatic enzyme levels without adverse effects on the fetus (1)(2)(3).

Fetal and maternal outcomes in women with PSC depend on the concurrent management of liver and bowel disease that may be independently associated with adverse pregnancy outcomes(3). Maternal outcome in PSC pregnancies is impaired. De novo pruritus and abdominal pain are the most frequently reported symptoms during pregnancy (4). Most women maintain stable serum liver test results. Notably, in a German cohort, 67% of pregnant women who received ursodeoxycholic acid (UDCA) had stable serum liver tests, compared to only 13% of those who did not receive UDCA. However, up to one-third of mothers experienced a decline in serum liver test results postpartum(5).

Ultrasound is the preferred initial imaging test. Magnetic resonance cholangiopancreatography (MRCP) is considered safe during pregnancy and can be used for diagnostic purposes. Depending on symptom severity, gestational stage, and the presence of significant bile duct strictures on imaging, endoscopic retrograde cholangiopancreatography (ERCP) can be performed for therapeutic bile duct interventions during the second or third trimester. Although radiation exposure is a concern in the

first trimester, ERCP may be considered in early pregnancy if the woman is acutely unwell, as the fetal radiation exposure remains well below the 50 mGy threshold associated with potential fetal risk (3).

Fetal outcomes in PSC pregnancies are compromised. A German cohort reported an early fetal loss rate of 16%, while preterm births occurred in 10% to 30% of cases (5). A Swedish study confirmed an increased preterm birth rate (16.3% vs. 5.1%), with approximately half being iatrogenic. The study also noted a higher rate of Caesarean deliveries (29.4% vs. 13.3%), independent of inflammatory bowel disease (IBD). However, no significant differences were observed in small-for-gestational-age births, stillbirths, congenital malformations, or neonatal deaths (6).

Conclusion:

Primary sclerosing cholangitis (PSC) occurring during pregnancy is rare. While PSC can impact both maternal and fetal health, the overall pregnancy outcome is generally favorable. Although UDCA often helps alleviate pruritus, there is limited data on its effectiveness in preventing stillbirth and its safety for the fetus or newborn.

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