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Case Report

Primary Sclerosing Cholangitis - An Unusual Presentation.

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Abstract

Introduction: Primary sclerosing cholangitis (PSC) is a chronic, progressive liver disease where bile ducts become inflamed, scarred, and narrowed, leading to bile buildup and liver damage. The exact cause is unknown, but it is strongly linked to inflammatory bowel disease (IBD) and is likely an autoimmune condition. Symptoms can include fatigue, itchy skin, and abdominal pain, and a liver transplant is often the only cure.
Case Report: We report a fourty year-old female, not a known case of any chronic illness and presented for first time with jaundice during second trimester of pregnancy and at that point of time was diagnosed to be having intrahepatic cholestasis of pregnancy. All her viral markers were negative. After delivery, jaundice did not completely subside and itching, though became less but persisted. She used to have intermittent exacerbation of jaundice which was associated with fever and chills. It used to subside after symptomatic treatment. These episodes remained for next few years but repeated ultrasonogram were reported to be normal. Later on, she developed pedal edema and at this point of time was diagnosed to be having changes of chronic liver disease and splenomegaly. She was put on standard treatment for chronic decompensated liver disease but despite that she progressed and developed persistent pancytopenia due to hypersplenism. She needed repeated blood transfusions for severe anemia and repeated injectable antibiotics for recurrent infection. Thus, on consultation with surgical team, she underwent splenectomy for hypersplenism. Her pancytopenia improved substantially and for the first time, ultrasonogram abdomen revealed dilated and irregular common bile duct. The MRCP done revealed multiple strictures and dilatation at level of intrahepatic ducts and dilated CBD with stricture at lower end, suggestive of primary sclerosing cholangitis. The liver biopsy also showed features of PSC. Her ursodeoxycholic acid has been increased to 25 mg/kg body weight, in addition to other symptomatic therapy including diuretics, multivitamin, calcium etc.
Conclusion: In pregnancy, as a body undergoes various physiological and hormonal changes, pregnant patients may present with symptoms of liver diseases, which normally subside after delivery. However, diagnosis and occurrence of PSC during pregnancy is quite rare but cannot be ignored. Therefore, critical attention shall be paid to pregnant women who report any fluctuations in liver enzymatic levels or complain of pruritis, as late diagnosis can lead to severe complications and maternal and fetal mortality.

Keywords: Primary Sclerosing Cholangitis, Cholangitis, Jaundice, Itching, Fatigue, MRCP

INTRODUCTION

Primary sclerosing cholangitis (PSC) causes chronic inflammation and scarring (sclerosis) of the bile ducts, which transport bile from the liver to the small intestine. This narrowing and scarring obstructs the flow of bile, causing it to back up and build up in the liver which eventually lead to cirrhosis and liver failure. The exact cause is unknown, but it is believed to be an autoimmune disease. It is strongly associated with inflammatory bowel disease, particularly ulcerative colitis; up to 80% of people with PSC also have IBD. Other potential factors include genetic predisposition and, possibly, exposure to certain bacteria or viruses. Many people are asymptomatic at diagnosis but when symptoms appear, they can include fatigue, abdominal pain, itching, jaundice, fever & chills and weight loss. The diagnosis depends upon blood investigations like—and imaging like MRCP and cholangiogram. The treatment of PSC is limited and focused on managing symptoms and complications. The only drug approved and has some beneficial effect is ursodeoxycholic acid. Liver transplantation is the only definitive cure and is required for many patients with end-stage liver disease. PSC is a risk factor for other cancers, including bile duct, colon, and gallbladder cancer.

CASE REPORT

We report a fourty year-old female, not a known case of any chronic illness and presented for first time with jaundice during second trimester of pregnancy and at that point of

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time was diagnosed to be having intrahepatic cholestasis of pregnancy. On biochemical evaluation she had microcytic hypochromic anemia on complete hemogram, liver function test was deranged i.e. there was low albumin level, mild hyperbilirubinemia, raised transaminitis with and mildly increased INR. The complete lipid profile, renal function test, serum electrolytes and blood sugar level were in normal range. All her viral markers were negative. After delivery, jaundice did not completely subside and itching, though became less but persisted. She used to have intermittent exacerbation of jaundice which was associated with fever and chills. It used to subside after symptomatic treatment. These episodes remained for next few years but repeated ultrasonogram were reported to be normal. Later on, she developed pedal edema and at this point of time was diagnosed to be having changes of chronic liver disease and splenomegaly. She was put on standard treatment for chronic decompensated liver disease but despite that she progressed and developed persistent pancytopenia due to hypersplenism. She needed repeated blood transfusions for severe anemia and repeated injectable antibiotics for recurrent infection. Thus, on consultation with surgical team, she underwent splenectomy for hypersplenism. Her pancytopenia improved substantially and for the first time, ultrasonogram abdomen revealed dilated and irregular common bile duct. The CECT scan abdomen showed altered echotexture of liver and multiple strictures and dilatation at level of intrahepatic ducts. The MRCP done revealed dilated CBD with multiple strictures and dilatations in intrahepatic ducts, more on right side, suggestive of PSC. The liver biopsy showed features of primary sclerosing cholangitis. The colonoscopy done to rule out inflammatory bowel disease and was found to be normal. Her ursodeoxycholic acid has been increased to 25 mg/kg body weight, in addition to other symptomatic therapy including diuretics, multivitamin, calcium etc.

Figure 1. Showing Dilated CBD with multiple strictures In both hepatic ducts, predominantly on right side.

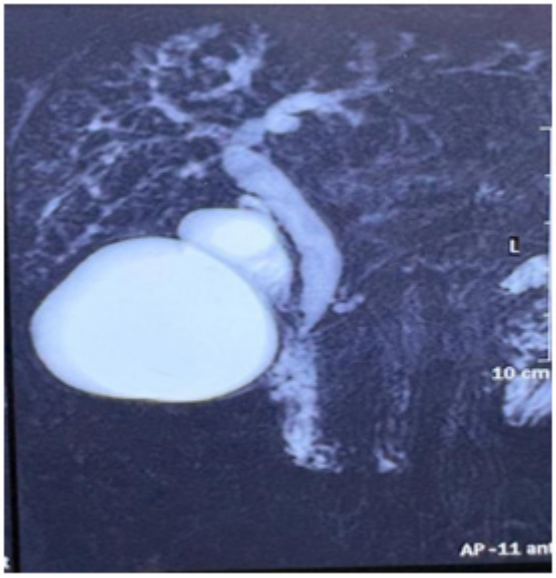


Figure 2. CECT Scan abdomen showing altered liver echotexture with multiple strictures in Intra hepatic ducts.

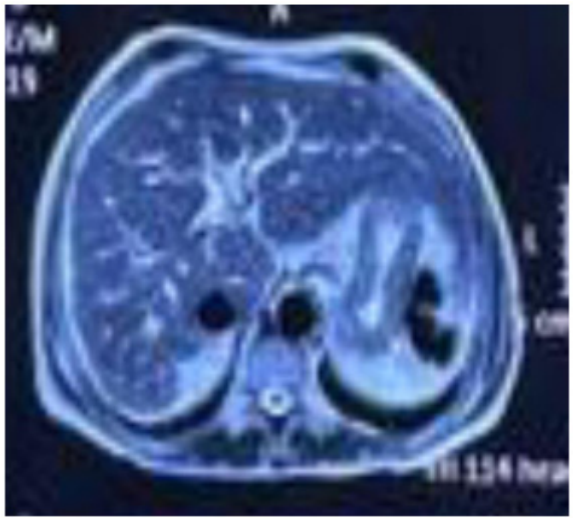
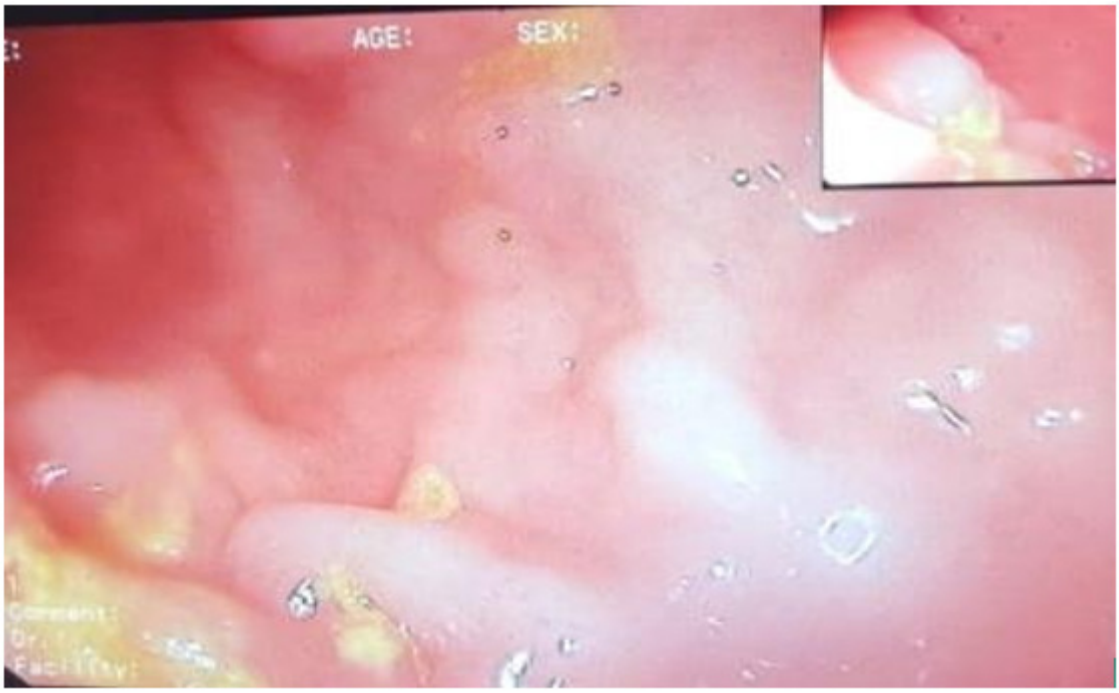


Figure 3. Showing normal colonoscopy in primary sclerosing cholangitis patient.



DISCUSSION

Liver disease imposes a considerable risk to both the mother and the fetus during pregnancy and may develop inadvertently during pregnancy. Primary sclerosing cholangitis (PSC) is a rare cause of liver disease in pregnancy [1]. PSC is an uncommon chronic cholestatic liver disease characterized by inflammatory damage of the intrahepatic and/or extrahepatic bile ducts, which results in bile stasis, fibrosis, and ultimately cirrhosis [2]. PSC is seldom identified for the first time during pregnancy. While pregnancy-related factors should be considered, the timing of symptoms plays a significant role in identifying the underlying cause. A multidisciplinary team is needed to manage PSC during pregnancy to maximize results for both the mother and the fetus [3]. MRCP can often be used to diagnose suspected PSC cases, negating the need for ERCP and its attendant risks [4]. Interval MRI/MRCP changes in the morphological appearance of the liver and biliary tree are frequently utilized to infer whether the illness is stable or has advanced [5]. PSC is usually seen in males but female can develop it, as in our case. In beginning PSC can be missed, especially in pregnancy where intrahepatic cholestasis of pregnancy (ICP) comes in mind first of all, same happened in our case. The other atypical presentation in our case was that usually PSC is diagnosed first and later on these patients develop chronic liver disease (CLD) but in our case, it was reverse and PSC was diagnosed much later after diagnosis of CLD, even after years of decompensation and splenectomy for persistent pancytopenia.

CONCLUSION

Primary sclerosing cholangitis is an uncommon cause of jaundice in pregnancy but should be kept in mind. In majority of pregnant patients who develop jaundice, intrahepatic cholestasis of pregnancy is thought but atypical causes like PSC can have first manifestation in pregnancy. This merits need of follow up of patients who were labelled as ICP during pregnancy.

Conflict Of Interest

The authors declare that there was no conflict of interest or any kind of funding was taken for publishing this case report.

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