Case Report

Open Access

CrossMark

Primary sclerosing cholangitis revealed by pregnancy

Abstract

Introduction: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease of poorly understood, probably dysimmune, mechanism. It is a slow-growing disease, with a tendency to worsen. We are reporting a rare case of primary sclerosing cholangitis revealed during pregnancy.

Case presentation: A 30-year-old woman who had pruritus during 12 weeks of gestation with a disturbance of the liver biologic tests. Imaging investigations found a chronic liver with areas of stenosis and dilation of VBIH and right and left hepatic bile ducts suggestive of primary sclerosing cholangitis with evidence of portal hypertension. She was put under treatment by ursodeoxycholic acid and the pregnancy proceeded without maternal-fetal complications giving birth to a healthy newborn with uneventful postpartum follow-up.

Conclusion: This case highlights the importance of considering pathologies other than benign intrahepatic cholestasis of pregnancy as the cause of cholestasis during pregnancy.

Keywords: pregnancy, cholangitis, cholestasis

Volume 14 Issue 2 - 2023

Alpha Boubacar Conte, Fatoumata Coulibaly, Sofia Jayi, Fatima Zohra Fdili Alaoui, Hikmat Chaara, Moulay Abdelilah Melhouf

Sidi Mohammed Ben Abdellah University, Department of Gynecology-Obstetrics II, Hassan II University Hospital, Fez, Morocco

Correspondence: Alpha Boubacar Conte, Resident of gynecology in the department of Gynecology-Obstetrics II, Hassan II University Hospital, Fez, Morocco, Email abccont33@yahoo.com

Received: March 29, 2023 | **Published:** April 11, 2023

Introduction

Primary sclerosing cholangitis (PSC) is a chronic cholestatic disease, characterized by inflammatory and fibrosing involvement of the bile ducts, of poorly understood, probably dysimmune mechanism.[1] It is a rare disease (prevalence of 10 to 40 per million) usually occurring in young men.[2] In women it can be revealed by pruritus during a pregnancy.[3] The possibility of underlying liver disease revealed by pregnancy should be considered, as it may have different prognostic implications for the mother and fetus. Thus, when cholestasis appears during pregnancy, it does not always involve the diagnosis of intrahepatic cholestasis of pregnancy (ICP), the most common cause of cholestasis during pregnancy.[4] We report in this article a case of PSC revealed during pregnancy. Through this case we did a brief review of the literature.

Patient and observation

Mrs. O.: aged 30 years, primiparous who consulted in the 2nd trimester of her pregnancy for an isolated and disabling generalized pruritus evolving since the 1st trimester (12weeks of gestation).

The first clinical examination found: a blood pressure of 11/06cmhg, negative urine test; temperature at 36.8°C with the presence of scratching lesions on the abdomen, both upper and lower limbs without obvious dermatological cause.

Laboratory tests revealed: hemoglobin 13.4 g/dl (12,5–15,5); platelet 164000/mm^3 (150000–400000); White blood cells 6720/mm^3 (4000–1000), ALAT 420UI/L (0–35) and ASAT 147UI/L (0–35), total bilirubin 72mg/l (3–12) and direct bilirubin at 4,8mg/l (0–2); alkaline phosphatases 192UI/L (30–120); ɤ-glutamyl transferase 76UI/L (0–38); prothrombin level at 100%; correct albumin. Bile acid testing returned to 48.4µmol /l (<8).

Cytobacteriological examination of urine and serologies (hepatitis C and cytomegalovirus) were negative.

The immunological assessment (anti-mitochondrial antibodies, anti-smooth muscle AC, anti-nuclear antibodies, anti-LKM1, anti-ALS) was negative.

Hepatobiliary ultrasound had objectified a chronic liver disease, without dilation of the bile ducts, without lithiasis. The imaging was supplemented by bili-MRI which highlighted a chronic liver with areas of stenosis and dilation of VBIH and right and left hepatic bile ducts suggesting primary sclerosing cholangitis with presence of signs of portal hypertension.

The other etiologies of sclerosing cholangitis having been eliminated, the diagnosis was primary sclerosing cholangitis in the stage of cirrhosis.

A screening Fibroscopy was performed showing esophageal spider veins.

In addition, in terms of obstetrics Fetal active movements were well perceived, there was no fetal heart rate abnormality and obstetric ultrasound objectified a progressive single-fetal pregnancy, a fluid in normal quantity with biometrics that corresponded to the 50-90th percentile of 29 weeks of amenorrhea.

She was put under ursodeoxycholic acid (AUDC) at the dose of 15mg/kg/day or 500mg/day initially with gradual increase of doses given cirrhosis up to 750mg/day.

The pregnancy was monitored in consultation with the hepato-gastrologist and was based on clinical (pruritus intensity), biological (liver assessment/2-3 weeks) and fetal monitoring (fetal active movement, fetal heart, and obstetric ultrasound).

The pregnancy proceeded without incident with a marked improvement in pruritus and liver tests. Follow-up intensified from the 37th week of gestation and consisted of bi-weekly fetal monitoring.

We performed the induction of the labor at 39 weeks of gestation. Mrs. O finally delivered by caesarean section following a failure of induction giving birth to a healthy newborn male with a weight of 3800g.

In addition, a colonoscopy performed postpartum did not find an association with inflammatory bowel disease.

Discussion

Several chronic liver diseases, sometimes previously asymptomatic, can, during pregnancy, cause cholestasis that should not be classified as “cholestasis gravidarum”. These diseases are chronic hepatitis C virus (HCV) infection; primary biliary cirrhosis and Primary sclerosing cholangitis.[3]

Primary sclerosing cholangitis (PSC), of undetermined cause, is a chronic cholestatic condition that results in irregular obliteration of the common bile duct, including the formation of multifocal structures. PSC is a progressive disorder that leads to liver cirrhosis and liver failure.[5,6]

The etiology of PSC is unknown, although the involvement of genetic susceptibility factors has been demonstrated.[7] It is associated in 70% of cases with inflammatory bowel disease (IBD) that often precedes PSC, five to ten years.[1] Typical symptoms of PSC include pruritus, abdominal pain in the right upper quadrant, fatigue, weight loss, and episodes of fever and chills.

Biologically, there is most often fluctuating cholestasis and transaminases are moderately increased.[2] Routine antibody testing is not necessary to diagnose PSC even though a wide variety of autoantibodies have been observed in PSC.[8] Analysis of anti-nuclear antibodies and anti-smooth muscle antibody may be appropriate for a subgroup of patients to confirm suspicions of "autoimmune" characteristics that may lead to therapeutic incidents.[6] The detection of biliary abnormalities remains the key element of diagnosis.

The reference examination is the direct opacification of the biliary tractus (endoscopic retrograde catheterization or cholangio-MRI)[9] as it was the case of our patient. The abnormalities observed are often long, sometimes multiple, typically without clear upstream dilation; a rosary aspect is very evocative. The involvement is most often intra- and extra-hepatic, rarely only intrahepatic (< 20%) which was the case of our patient or only extra-hepatic (< 10%).[9]

In case of biological cholestasis associated with bile duct irregularity with biliary structures-dilatations on bili-MRI, liver biopsy is not necessary for diagnosis. It will be performed in case of normal MRI, or if presence of antinuclear antibodies, anti-smooth muscle, increased IgG or transaminases suspecting associated autoimmune hepatitis.[2]

Given the frequent association with IBD, total colonoscopy combined with biopsies should be performed in patients diagnosed with PSC in the absence of known IBD.[6] In case of pregnancy in a woman with PSC, the risks of prematurity and caesarean section are increased, but not those of congenital malformation.[10]

Several studies show that maternal complications do not increase in patients with PBC during pregnancy.[11–13]

Today, there is no standard treatment for PSC. Therapeutic trials with ursodeoxycholic acid (UDCA) have shown that it improves pruritus as well as biological parameters.[1] We also note a clear improvement in the clinical and biological symptomatology under UDCA treatment in our patient AUDC can be safely used during pregnancy in patients with PBPC.[11]

Conclusion

Pregnancy by altering hepatic metabolism may be the revealing way of underlying chronic liver disease that should not be mistaken as cholestasis of pregnancy. Given the risks that patients of primary sclerosing cholangitis run subsequently (cirrhosis, IHC, IBD), it is essential to diagnose them to consider an adequate follow-up.

Acknowledgments

None.

Ethics approval and consent to participate: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials: All the data available are presented in this report.

Authors' contributions: All the authors participated in the management of the patient and the writing of the manuscript. All authors read and approved the final manuscript.

Funding

We did not receive any funding for the study.

Conflicts of interest

The authors declare that they have no competing interests.

References

1. Émile C. Adult cholestatic diseases: primary biliary cholangitis and primary sclerosing cholangitis. *Option/Bio*. 2021;32(631):21–22.
2. Mallet V, Corouge M, Sogni P. Liver and autoimmunity. *Rev Francoph Lab*. 2013;449:30–34.
3. Audrey P, Arnaud B, Teodora R, et al. Liver and pregnancy. Obstétrique pour le praticien 7 IE.
4. Dixon PH, Williamson C. The pathophysiology of intrahepatic cholestasis of pregnancy. *Clin Res Hepatol Gastroenterol*. 2016;40(2):141–153.
5. Lee RH, Goodwin TM, Greenspoon J, et al. The prevalence of intrahepatic cholestasis of pregnancy in a primarily Latina Los Angeles population. *J Perinatol*. 2006;26(9).
6. European Association for the Study of the Liver. EASL Clinical Practice Guidelines: Management of cholestatic liver diseases. *J Hepatol*. 2009;519(2):237–267.
7. Karlsen TH, Schrumpf E, Boberg KM. Genetic epidemiology of primary sclerosing cholangitis. *World J Gastroenterol*. 2007;13(41):5421–5431.
8. Hov JR, Boberg KM, Karlsen TH. Autoantibodies in primary sclerosing cholangitis. *World J Gastroenterol*. 2008;14(24):3781–3791.
9. Chazouilléres O. Biology days maladies cholestatics chronicles day La' Adulthe. *Rev Francoph Lab*. 2007;394:25–32.
10. Ludvigsson JF, Bergquist A, Ajne G, et al. A population-based cohort study of pregnancy outcomes among women with primary sclerosing cholangitis. *Clin Gastroenterol Hepatol Off Clin Pract J Am Gastroenterol Assoc*. 2014;12(1):95–100.
11. Arora A, Kumar A, Anand AC, et al. Indian national association for the study of the liver—federation of obstetric and gynaecological societies of India position statement on management of liver diseases in pregnancy. *J Clin Exp Hepatol*. 2019;9(3):383–406.
12. Ferrigno B, Barba R, Medina-Morales E, et al. Cholestatic liver disease and pregnancy: a systematic review and meta-analysis. *J Clin Med*. 2022;11(4):1068.
13. Wellge BE, Barba R, Esli MM, et al. Pregnancy in primary sclerosing cholangitis. *J Clin Med*. 2011;60(8):1117–1121.