

A possible overlap between acute fatty liver of pregnancy and intra hepatic cholestasis of pregnancy: Case report

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Abstract

Liver diseases that are unique to pregnancy are usually classified based on timing of presentation in relation to gestational age, among those conditions, intrahepatic cholestasis of pregnancy (ICP), and acute fatty liver of pregnancy (AFLP) typically occur during the second trimester and should be differentiated as management is different. Liver biopsy can help distinguish between those two conditions, however, the invasive nature of the biopsy makes it a hard decision to proceed with in clinical practice. We present a case report of a pregnant patient with possible overlap between ICP and AFLP, for which liver biopsy dictated the management.

Keywords

intrahepatic cholestasis of pregnancy; acute fatty liver of pregnancy; liver disease in pregnancy.

Introduction

Liver diseases that are unique to pregnancy are usually classified based on timing of presentation in relation to gestational age, which includes hyperemesis gravidarum, preeclampsia/eclampsia; the syndrome of hemolysis, elevated liver enzymes, and low platelets, intrahepatic cholestasis of pregnancy (ICP), and acute fatty liver of pregnancy (AFLP). Among those conditions, ICP and AFLP typically occur during the second trimester [1]. Both ICP and AFLP have different pathophysiology, unique way of presentation, and different treatment strategy, and it is important to differentiate between both conditions since AFLP can be life threatening and can lead to acute liver failure [2]. We describe a case with possible overlap between ICP and AFLP.

Case Presentation

A 39-year-old female, 24-weeks pregnant, presented with nausea, vomiting, fatigue, and pruritus. She had no previous history of liver disease. Physical examination showed icterus, normal liver size and no other signs of chronic liver disease. Serological workup showed ALT 1274, AST 1204, total bilirubin (TB)6.9, alkaline phosphatase (ALP)183. Workup included serological workup for viral hepatitis (includes Hepatitis C), autoimmune hepatitis, ultrasound of the liver, and uric acid, and all were normal except for elevated bile acid of 156. Given likely diagnosis of ICP, she was given supportive care and initiated on Ursodeoxycholic acid (UDCA) and discharged home after slight improvement. A week later, patient was readmitted due to worsening pruritus and worsening liver function test. Her TB was 15, bile acid level 374, and INR of 2.5 (Table 1). This time, a liver biopsy was obtained and sent to an expert pathologist in liver diseases. H&E stained sections showed “lobular disarray with microvesicular steatosis and necrotic hepatocytes concerning for AFLP” (Image 1). Unfortunately, Oilred O stain was not performed as the sample was fixed before it was sent out. Soon after the final pathology report, the patient underwent emergent caesarian section at 27 weeks with significant improvement in the TB, INR and bile acids level few days after. Table 1 shows the laboratory values at different times during the clinical course.

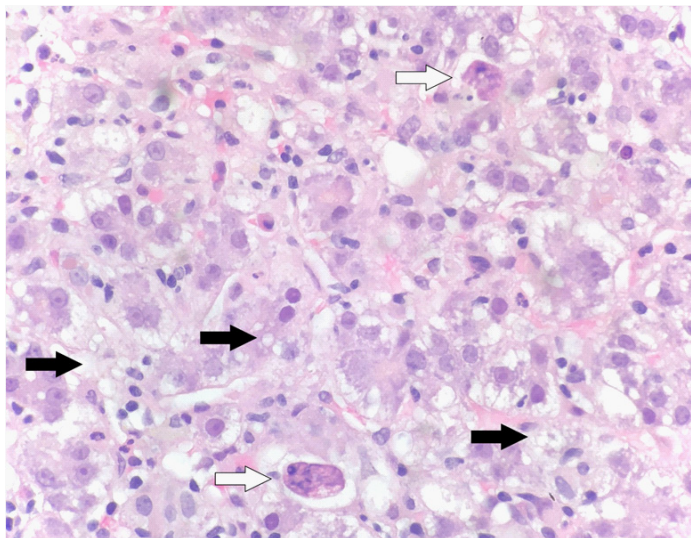


Figure 1: Demonstrates H&E stained sections which shows lobular disarray with microvesicular steatosis (black arrows) and necrotic hepatocytes (white arrows).

Discussion

Evaluation and management of liver disease in pregnancy is unique and challenging. In addition to the normal workup that we perform for non-pregnant patients, one has to think about liver disorders related to pregnancy itself, including hypertensive disorders of pregnancy, AFLP, and ICP. ICP and AFLP have different presentation clinically and laboratory that is imperative to distinguish given how serious AFLP can be³. ICP, which is the most common, typically presents in the second or third trimester. Main symptom is pruritus, but also fatigue, anorexia, and jaundice could happen. The typical laboratory finding in this condition is elevated bile acids, which would be concerning for complications if level exceeds 40 Mmol/L⁴. These complications include still birth, meconium aspiration, respiratory distress syndrome and asphyxia.

Table 1: Shows the laboratory values during the disease course.

	First admission	First discharge	Second admission	Pre-delivery	Post- delivery	2 weeks later	Normal range
Platelets 10*3/uL	260	237	397	311	316	168	140-440
AST U/L	1036	1204	855	551	333	131	15-46
ALT U/L	1125	1274	748	371	424	170	0-35
Total Bilirubin mg/dL	5.7	6.9	15.8	11.9	11.5	3.9	0.2-2.3
Alkaline Phosphatase U/L	180	183	222	158	302	253	38-126
Bile acids	156	157	374	363	169	68	0-10
INR	1.2	1.2	1.4	2.5	1.3	1.3	1.0-1.2

Treatment recommendations for ICP include UDCA which shown to reduce pruritus, ALT and bile acid levels; and appropriate planning for delivery around 37 weeks². On the other hand, Acute Fatty liver of pregnancy typically presents in the third trimester between 28-36 weeks with abdominal pain, nausea, vomiting, fatigue, anorexia, and often encephalopathy if it progresses to acute liver failure (ALF). AST and ALT elevation are similar to ICP with levels up to 20 times the upper limit of normal [2]. A clue to the diagnosis would be fatty liver on imaging studies. The Swansea criteria have been proposed as a diagnostic tool for AFLP⁵. Once diagnosed, it is an obstetric emergency that warrants prompt delivery of the fetus, given high maternal mortality of 10-20%, and fetal mortality up to 20% [5]. Both mothers and fetuses require monitoring post-partum given the risk of ALF. Those patients may benefit from genetic testing as AFLP may recur in subsequent pregnancies. Liver biopsy can help distinguish between the above conditions [4]. However, the invasive nature of the biopsy makes it a hard decision to proceed with. It is imperative to understand the differences and distinguish between ICP and AFLP given their implications on the mother and fetus. The case we report presented initially as ICP given pruritis, cholestatic pattern of liver enzymes elevation, elevation of bile acid, and normal appearing liver on ultrasound, however, the liver biopsy suggested a pattern consistent with acute fatty liver of pregnancy which may suggest a possible overlap between the two disorders in this case. The lesson we learned from this case is to always consider preparing a frozen tissue for Oilred O stain whenever a liver biopsy has been planned for a pregnant patient with abnormal liver enzymes.

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Manuscript Information: Received: October 12, 2021; Accepted: December 20, 2021; Published: December 30, 2021

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Citation: Eisa M, Safadi S, Gondim DD, Marsano L. A possible overlap between acute fatty liver of pregnancy and intra hepatic cholestasis of pregnancy: Case report. *Open J Clin Med Case Rep.* 2021; 1820.

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