Acute fatty liver of pregnancy accompanied with disseminated intravascular coagulopathy and encephalopathy: a case report

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Abstract

Acute fatty liver disease of pregnancy (AFLP) is a rare condition associated with other common liver manifestations such as hemolysis, elevated liver enzymes, and low platelets syndrome (HELLP). We present a 27-year-old pregnant woman who developed hepatic encephalopathy and DIC after being diagnosed with Acute fatty liver disease of pregnancy.

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Consent statement:

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Abstract

Acute fatty liver disease of pregnancy (AFLP) is a rare condition associated with other common liver manifestations such as hemolysis, elevated liver enzymes, and low platelets syndrome (HELLP). We present a 27-year-old pregnant woman who developed hepatic encephalopathy and DIC after being diagnosed with Acute fatty liver disease of pregnancy.

Key Clinical Message:

When common causes are ruled out, a pregnant woman with a fever, low platelets, and abnormal liver function should be evaluated for Acute fatty liver disease of pregnancy with a thorough history, detailed examination, and proper investigation, and treatment should begin as soon as possible to avoid potentially fatal complications.

Keywords: Fatty Liver, Jaundice, Hepatology, Fetus, Obstetrics, Pathology.

Abbreviations: AFLP = Acute fatty liver disease of pregnancy, HELLP = Hemolysis, Elevated Liver Enzymes and Low Platelets Syndrome, LFTs = Liver Function Tests, DIC = Disseminated Intravascular Coagulopathy, ICU = Intensive Care Unit

Introduction

Acute fatty liver disease of pregnancy (AFLP) is an uncommon condition compared to other causes of pathologies in the liver such as hemolysis, elevated liver enzymes, and low platelets (HELLP) syndrome.[1] It is a life-challenging disease that has a high prevalence of about 1 to 3 cases per 10,000 deliveries occurring mostly at 36 weeks of gestation marked by fatty infiltration of the liver leading to hepatic failure.[2] The management of AFLP is prompt delivery of a fetus with improvements being noticed after 1-2 days of delivery.[3] Early diagnosis is necessary to ensure better maternal and fetal outcomes.[4] Clinical findings in AFLP can vary, and diagnosis is further complicated due to significant overlap with the clinical and biochemical markers of HELLP syndrome.[5] The main finding related to AFLP includes infiltration of hepatocytes with fats including the pericentral zone while sparing the periportal hepatocyte.[6] A higher rate of mortality is observed in developing countries due to a shortage of capacity for intensive care. If AFLP is left untreated then jaundice can begin abruptly causing serious damage further progressing to fulminant hepatic failure and its associated complications.[7] This is the stage where most of the AFLP is diagnosed.

To our knowledge, there is no case yet reported with the delivery of an alive fetus in a patient with AFLP at 26 weeks of gestation. Hence, we are presenting a case of a pregnant 27-year-old female with AFLP. She was delivered an alive male fetus due to prompt and early diagnosis despite being manifested with hepatic encephalopathy and disseminated intravascular coagulopathy (DIC).

Case presentation

A 27-year-old woman (G3 P2+0), with three confirmed pregnancies and two previous deliveries, presented at 26 weeks of gestation in a tertiary care hospital with a history of fever, low platelets, and deranged liver function tests (LFTs) for two days. Her pregnancy was un-booked, and the above-mentioned findings were detected at a local clinic. Before this presentation, the patient was not on any medications.

On presentation to the hospital, physical examination revealed a well-nourished and conscious woman who was oriented to time, place, and person. Her pulse rate was 82 beat per minute, blood pressure 120/80 mmHg, and oxygen saturation of 95% at room air. Abdominal examination revealed a height of the fundus corresponding to 26cm. Neurological examination was non-significant and the Glasgow Coma Scale (GCS) was 15/15. The rest of the physical examination was unremarkable.

She was started on empiric antibiotics and further investigations were done including Dengue NS1 antigen, LFTs, and Urine DR, to rule out other causes of her symptoms. A few hours into admission, the patient developed hypoglycemia for which she was intravenously given 25% dextrose.

A chest x-ray was performed showing right pleural effusion and bilateral atelectasis. Ultrasound of the abdomen confirmed fatty changes with a normal-sized hypoechoic liver sparing periportal zones ruling out

HELLP (Figure 1). A provisional diagnosis of AFLP (and viral hepatitis) was made.

A normal sonogram of the gall bladder, pancreas, spleen, kidney, and urinary bladder was reported. The surface Antigen test showed no reaction to HbsAg and HCV antigen, so ruling out viral hepatitis. A confirmed diagnosis of AFLP was made.

12 hours into admission, the patient complained of abdominal pain, vomiting, constipation, and headache, for which she was promptly transferred to the intensive care unit (ICU). The patient developed significant dyspnea with a total leukocyte count of 24.26 x 109/L, severe anemia, and a left-sided consolidation on CXR.

She developed tachycardia with a pulse of 118/bpm. Fetal movements were positive. Her hemoglobin level dropped from 11.3g/dL to 8.6g/dL, with a prolonged APTT and INR, and her serum SGPT had risen to 3339 IU/L recorded in Table 1. The patient became markedly drowsy, irritable, and restless, and a diagnosis of Grade 1 (hepatic) encephalopathy was made, for which she was given lactulose. Later patient progressed to Grade II hepatic encephalopathy, with a GCS of 13/15.

Vaginal delivery was chosen over a cesarean to prevent further deterioration of her condition. An alive male fetus was delivered, with a poor APGAR score and was handed for resuscitation. Around 600-700ml of clots were removed from the uterus.

An hour later, fresh bleeding was observed per vaginal examination. With a contracted uterus, her perineum was inspected for tears, balloon tamponade was performed. However, the patient continued to bleed and was shifted to the operation theatre for uterine artery embolization. Her BP was 120/ 83 mmHg, pulse 120 bpm, and O2 saturation of 93% at room air. She was on continuous positive airway pressure (CPAP), and mannitol was administered to protect renal function after the vascular procedure.

The procedure successfully stopped her bleeding. The patient was drowsy, irritable, and not following commands, consistent with a progression to grade III hepatic encephalopathy, with a GCS of 11/15. Antibiotic treatment was continued.

Over the course of the next two days, the patient's condition improved steadily. CXR showed improvement and CT brain showed no cerebral edema (Figure 2).

Approximately two months later, the patient returned to our facility with a complaint of lower abdominal pain. On examination, her uterus was bulky, with a thickened endometrium and necrotic debris sloughing off through the vagina. She had developed uterine necrosis; a late and rare complication of the uterine artery embolization that had been performed for her postpartum hemorrhage. As a result, a total abdominal hysterectomy was performed, after which she was discharged from the hospital.

Discussion

Jaundice during pregnancy has many causes including but not limited to intrahepatic cholestasis, cholelithiasis, viral hepatitis, pre-eclampsia with or without HELLP syndrome, and AFLP. Intrahepatic cholestasis of pregnancy (ICP) may present during the third trimester but it is mainly characterized by pruritis in the absence of a skin rash with abnormal LFTs (elevated transaminase and bile acids) and usually resolves after birth, however, serum bilirubin concentration is rarely higher than 6mg/dl. Cholelithiasis may occur at any time during pregnancy and is accompanied by pain in the right upper quadrant, and fever, and ultrasonography (USG) is usually diagnostic.[8] Acute viral hepatitis in pregnancy presents as a systemic illness with fever, nausea, vomiting, fatigue, jaundice, dark urine and pale stools , and markedly elevated aminotransferase concentrations (>500U/liter).

The incidence of HELLP syndrome is three times as much as that of AFLP.[9] The symptoms of our patient were initially indicative of both HELLP and AFLP; nausea, vomiting, dyspnea, epigastric abdominal pain, anorexia , jaundice, and hence supportive treatment for AFLP and HELLP Syndrome was started. Further laboratory investigations indicated severe coagulopathy, raised APTT, elevated serum transaminase and bilirubin levels, hypoglycemia, an elevated ammonia value, and a low albumin level, favoring the diagnosis of AFLP over HELLP syndrome.

In AFLP there is an inherited enzyme deficiency in beta-oxidation, due to a defect in the enzyme LCHAD (long-chain hydroxy acyl-coenzyme A dehydrogenase) predisposing them to AFLP resulting in progressive lipid accumulation within the hepatocytes. However, other risk factors can also be noted e.g., primigravida (first pregnancy), pre-eclampsia, male fetus, and multiple gestation.[10]

Lastly, extrahepatic complications delayed maternal recovery for up to four weeks after delivery. These trends in earlier studies make this case report noteworthy as our patient not only presented late to us after the development of hepatic encephalopathy, acute renal failure, and DIC but also improved a week before delivery.

Conclusion

Early diagnosis and prompt management are essential to prevent maternal and perinatal morbidity and mortality. AFLP can be managed with early and prompt diagnosis through laboratory tests to reduce complications. Further studies can be conducted to devise a relationship between factors that can lead to termination of the pregnancy in patients with hepatic encephalopathy and DIC so neonatal deaths can be reduced and a solution can be stamped with regards to AFLP.

Competing Interests

None declared.

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Consent for publication

An institutional approval letter was granted by Ziauddin Medical University, Karachi, Pakistan for this case report.

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DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available with the corresponding author upon reasonable request.

Authors' contributions

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