Acute Fatty Liver of Pregnancy with Atypical Presentation: A Case Complicated by Multiorgan Failure and Severe Acute Pancreatitis

Abstract

Acute fatty liver of pregnancy (AFLP) is a rare but severe complication of pregnancy, typically occurring in the third trimester. Its diagnosis can be challenging due to its polymorphic clinical presentation, which may mimic other hepatic and systemic pathologies. We report the case of a 35-year-old primigravida patient who was initially hospitalized for acute non-lithiasic pancreatitis. Her condition rapidly deteriorated into multiorgan failure, with fulminant hepatitis, hepatic encephalopathy, and refractory septic shock, leading to a non-recoverable cardiac arrest despite intensive medical management.

Introduction

Acute fatty liver of pregnancy (AFLP) is a rare pregnancy-related condition, with an estimated incidence of 1 in 7,000 to 1 in 15,000 pregnancies. It usually occurs between the 30th and 38th weeks of gestation and is characterized by microvesicular hepatic infiltration, leading to acute hepatocellular failure. If not diagnosed and treated promptly, AFLP can progress to multiorgan failure, posing a significant risk to both maternal and fetal survival.

In this context, we present an atypical case of AFLP complicated by severe acute pancreatitis. This unusual association led to fulminant hepatic failure and septic shock, highlighting the diagnostic and therapeutic challenges of this condition.

Case Report

A 35-year-old primigravida (G1P0) with no significant medical history was admitted to the gynecology and obstetrics department (GO1) for acute non-lithiasic pancreatitis, diagnosed by MRI and

classified as stage B according to the Balthazar classification. She presented with intense epigastric pain and vomiting, with elevated lipasemia (501 mg/L) and CRP (96 mg/L). Obstetric ultrasound was reassuring, showing normal fetal growth and an adequate amount of amniotic fluid. After symptomatic management, she was discharged with a scheduled outpatient follow-up.

One week later, the patient was readmitted in an emergency setting with altered consciousness associated with severe hypoglycemia (0.23 g/L). On clinical examination, she was conscious but slow to respond, with tachycardia at 135 bpm and blood pressure at 110/60 mmHg. She was also anuric and tachypneic, with an oxygen saturation of 92-94%. Laboratory findings revealed severe hyperlactatemia (11.6 mmol/L), hepatic cytolysis (AST 84N, ALT 23N), coagulopathy with a significantly reduced prothrombin time (8%), and acute renal failure (creatinine 26 mg/L, urea 0.52 g/L).

Obstetric ultrasound revealed intrauterine fetal death (IUFD) at 34 weeks of gestation, without signs of retroplacental hematoma. A thoraco-abdominopelvic CT scan showed severe pancreatitis (Balthazar E, severity score II) with infected necrotic collections, bilateral cortical renal necrosis, aseptic myomatous necrobiosis, and pleural and intraperitoneal effusions. The immediate evolution was marked by hemodynamic failure with hypotension (85/40 mmHg), persistent anuria, and septic shock requiring admission to intensive care.

In the intensive care unit, the patient was intubated, mechanically ventilated, and placed on norepinephrine. Broad-spectrum antibiotic therapy (imipenem and levofloxacin) was initiated. The diagnosis of AFLP complicated by fulminant hepatic failure secondary to IUFD was confirmed by the intensive care team. An attempt at labor induction using misoprostol (Cytotec®) was performed but was unsuccessful. Laboratory tests showed worsening liver function (AST 1242 IU/L, ALT 675 IU/L, prothrombin time 30%, total bilirubin 62 mg/L). Respiratory deterioration occurred, with acute respiratory distress syndrome (ARDS) and nosocomial pulmonary infection (PCR multiplex identified Acinetobacter baumannii, Haemophilus influenzae, and Staphylococcus aureus).

Due to the failure of labor induction, an emergency cesarean section was performed. The procedure required bilateral hypogastric artery ligation and B-Lynch sutures due to severe uterine atony. Despite optimal management, the patient's condition remained critical, with refractory septic shock progressing to non-recoverable cardiac arrest.

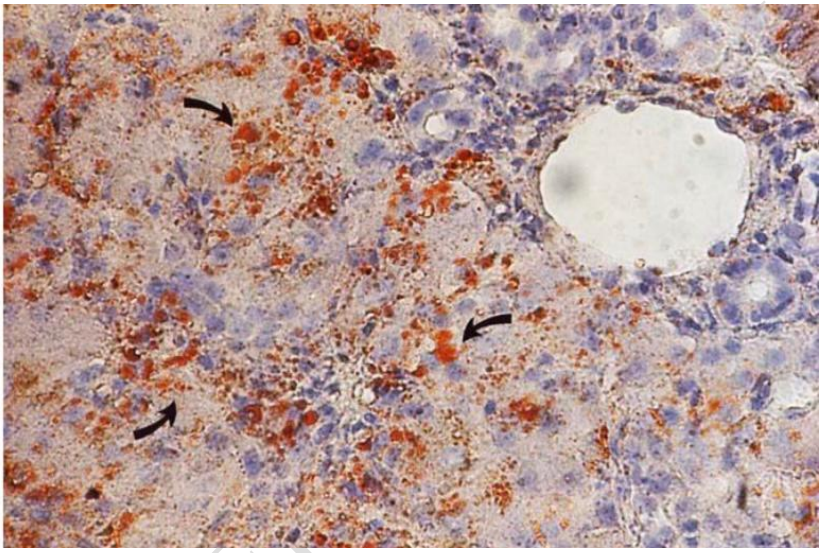

Figure 1: Liver biopsy (SHAG) with specific fat staining (Sudan IV) examined under a light microscope, showing fat droplets (black arrows) diffusely dispersed throughout the liver. [7]

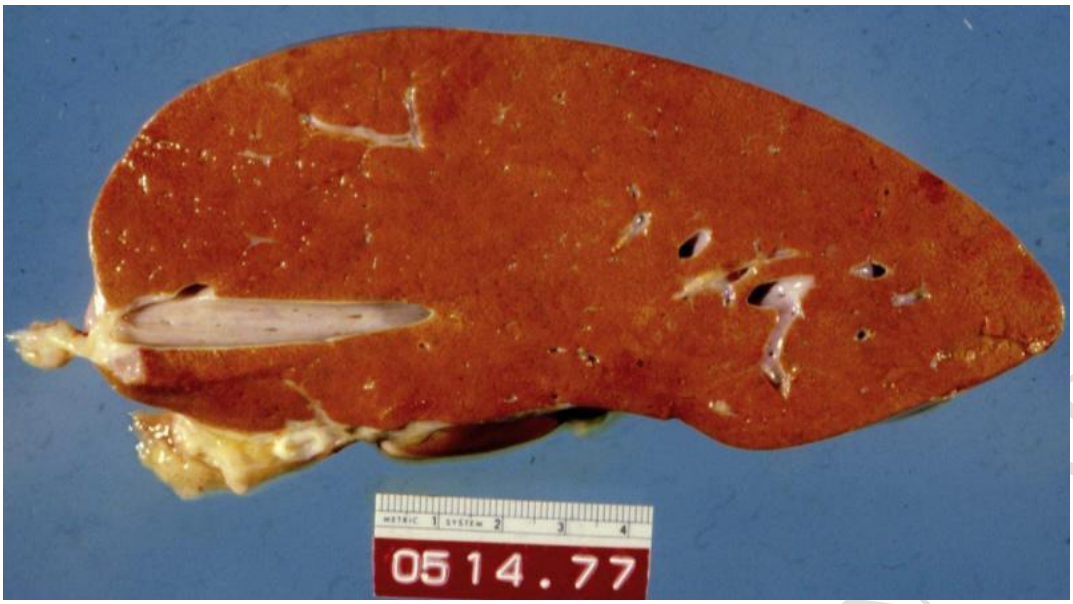

Figure 2: Autopsy specimen of a woman who succumbed to acute fatty liver of pregnancy. The liver exhibits a yellowish and fatty appearance. [8]

Discussion

AFLP is an obstetric emergency whose diagnosis is often delayed due to its variable clinical presentation and overlap with other pregnancy-related hepatic disorders such as HELLP syndrome, severe preeclampsia, or acute pancreatitis. It is caused by a mitochondrial enzyme deficiency (LCHAD) leading to triglyceride accumulation within hepatocytes and subsequent hepatocellular dysfunction.

Warning signs include unexpected hypoglycemia, acute hepatic failure with cytolysis and coagulopathy, and multiorgan dysfunction. In this case, the association with severe acute pancreatitis complicated the diagnosis and delayed the specific management of AFLP.

Treatment relies on the rapid delivery of the fetus, which remains the only curative therapy. Intensive supportive care is essential, including the correction of metabolic disturbances, hemodynamic stabilization, and close monitoring for infectious and hemorrhagic complications. Despite these measures, maternal prognosis remains poor in cases of late diagnosis, as illustrated by this case.

Conclusion

This case highlights the diagnostic complexity and severity of AFLP, particularly when associated with severe acute pancreatitis. Early diagnosis and aggressive management are crucial in reducing maternal and neonatal mortality. This case underscores the importance of heightened vigilance in the presence of unexplained hepatic abnormalities during pregnancy to prevent a fatal outcome.

References

1. Knight M, et al. “Maternal mortality from acute fatty liver of pregnancy.” Obstetrics & Gynecology, 2021.
2. Ch’ng CL, et al. “A clinical update on AFLP: Pathophysiology and management.” Liver International, 2020.
3. Riely CA, “Acute fatty liver of pregnancy.” Hepatology, 2019.
4. Sibai BM, et al. “Differentiating AFLP from HELLP syndrome.” American Journal of Obstetrics and Gynecology, 2018.
5. Vigil-De Gracia P, “Liver disorders in pregnancy.” Best Practice & Research Clinical Obstetrics & Gynaecology, 2019.
6. Tran TT, et al. “Management of liver diseases in pregnancy.” World Journal of Gastroenterology, 2020.
7. Humberto Reyes, ACUTE FATTY LIVER OF PREGNANCY, A Cryptic Disease Threatening Mother and Child. 1089-3261/99.
8. Nelson DB, Yost NP, Cunningham FG. Acute fatty liver of pregnancy: clinical outcomes and expected duration of recovery. Am J Obstet Gynecol 2013;209:1-7.456.