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An Unpredictable Course of Acute Fatty Liver of Pregnancy

ABSTRACT

Acute Fatty Liver of Pregnancy is an obstetric emergency associated with high maternal mortality and morbidity. A decade ago , the maternal mortality rates were significantly higher reaching upto 70 %. Presently maternal mortality has decreased to around 18% with associated neonatal mortality ranging from 7% to 66% attributed to focused liver intensive careand a robust multidisciplinary team approach.Our patient had a late and critical presentation with hepatic encephalopathy on admission. Swift and promt decisions with a good liver team backup can reduce the mortalty rates associated with AFLP. The patient had multiple life threatening catastrophic and turbulent events post Caesarean section. After weeks of intensive post operative care she was discharged from the hospital .We had a successful outcome inspite of delayed presentation .

Key words: AFLP, Pregnancy, Fatty Liver

INTRODUCTION

Acute fatty liver of pregnancy (AFLP) is an obstetric emergency associated with high mortality and morbidity. A decade ago, maternal mortality rates were significantly higher, reaching up to 70%. Presently, maternal mortality has decreased to approximately 18%, with associated neonatal mortality ranging from 7% to 66%, attributed to focused liver intensive care and a robust multidisciplinary team approach. [(1)]

AFLP is characterized by microvesicular steatosis in the liver, typically occurring in the third trimester of pregnancy. It is believed to result from mitochondrial dysfunction impairing fatty acid oxidation, leading to hepatocyte accumulation.[(2)] This condition can progress to acute liver failure, disseminated intravascular coagulation, postpartum hemorrhage, acute kidney injury, gastrointestinal bleeding, and, rarely, hepatic rupture. The primary treatment for AFLP is early termination of pregnancy to prioritize maternal safety.

CASE REPORT

Here, we discuss a c.ase of AFLP presenting in the third trimester with fulminant hepatic failure, necessitating an early cesarean section, followed by an unexpected tumultuous postoperative course.

A 27-year-old primigravida at 32 weeks gestation (last menstrual period 17/04/2023) was admitted with complaints of yellowish discoloration of the skin and eyes, vomiting, and fatigue persisting for 7 days, along with abnormal liver function tests. She was referred from a peripheral hospital in Maharashtra due to altered sensorium and worsening hemodynamic status.

On admission, she presented with hypotension, oliguria, severe metabolic acidosis (pH 7.16), an INR of 8.8, and an arterial ammonia level of 99 umol/L, indicating grade II hepatic encephalopathy. According to Swansea criteria, AFLP was suspected due to severe metabolic acidosis and hepatic encephalopathy.[(3)] Abdominal and pelvic ultrasound revealed grade 1 fatty liver infiltration, minimal left pleural effusion, and a single live intrauterine pregnancy at 33 weeks 1 day with breech presentation and reduced amniotic fluid (mild to moderate oligohydramnios, AFI 5cm).The Doppler finding was suggestive of placental insufficiency.

Due to worsening hepatic encephalopathy, she was electively intubated. Resuscitative efforts included crystalloid administration, N-acetylcysteine, sodium bicarbonate infusion, low-dose noradrenaline, and broad-spectrum antibiotics. Preoperatively, she received 2 units of fresh frozen plasma (FFP) and 10 units of cryoprecipitate based on thromboelastogram findings before being taken to the operating room.

A cesarean section was performed due to a high floating head and the patient's critical condition. A premature male infant weighing 1.6 kg was delivered with depressed respiration and a poor APGAR score, requiring Neonatal ICU admission.

Intraoperative estimated blood loss was 300 ml.Parient was transferred to the liver ICU, where she received 10 units of cryoprecipitate, 1 unit of single-donor platelets, and 2 units of FFP,due to severe coagulopathy and slightly hemorrhagic drainage output. Given persistent systemic inflammatory

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response syndrome (SIRS) and severe coagulopathy, a single session of therapeutic plasma exchange (TPE) with standard volume plasma (3.5 liters) was performed.

Subsequently, the patient showed clinical improvement with enhanced encephalopathy grade, increased urine output, and normalized blood pressure. Metabolic acidosis also improved with fluid resuscitation. However, approximately 20 hours postoperatively, she developed sudden tachycardia, hypotension, and significant hemorrhagic drainage (approximately 700 ml). Contrast-enhanced CT of the abdomen and pelvis confirmed bilateral rectus sheath hematoma, loculated ascites, and mild bilateral pleural effusion (Figures 1 and 2).

Figure 1 and 2 : Bilateral rectus sheath haematoma ascites and bilateral mild pleural effusion.

During re-exploration, a retro-rectus hematoma (approximately 400 ml) and pelvic clots (more than 200 ml) were noted with diffuse oozing in the rectus muscle. There was no active bleeding from the uterine closure site or angles. Hemoglobin dropped to 3.8 g/dl during surgery, necessitating intraoperative transfusion of four units of blood and one unit of single-donor platelets. Following the procedure, she was returned to the ICU with increased inotropic support.

Over the next 48 hours, the patient demonstrated neurological and hemodynamic improvement. However, due to reduced urine output, diuretics were initiated along with albumin infusion. After two days, her sensorium worsened with elevated blood ammonia levels, prompting reintubation and sedation for airway protection. Lactulose enemas were initiated, neurology consultation was obtained, and a normal CT brain was confirmed.

Given her deteriorating clinical condition and laboratory parameters, a transjugular liver biopsy revealed features consistent with acute fatty liver of pregnancy, without evidence of fibrosis or cirrhosis (Figure 3). With comprehensive supportive care and close monitoring, her condition gradually improved, and she was extubated within seven days. Following discharge from the ICU after 24 days of hospitalization, she transitioned to the ward with an abdominal drain. Initially, the drain output was 3.8 liters, decreasing to 1 liter with initiation of a high-protein diet and aggressive physiotherapy. Regrettably, her premature infant succumbed to poor general condition and premature delivery.

This critical case, characterized by an initial INR of 8.89 and a challenging postoperative course, underscores the efficacy of expedited delivery and a skilled intensive liver care team in achieving survival outcomes.

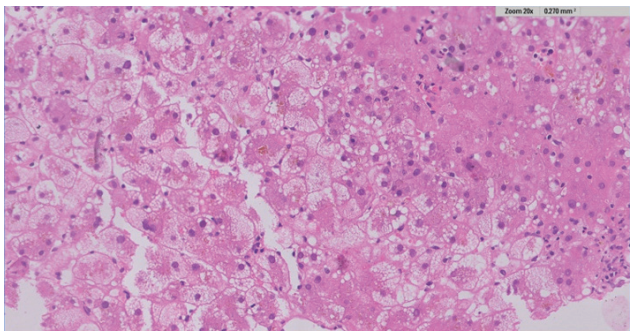

Figure 3: Histopathology of Liver tissue suggestive of severe necrosis, no evidence of fibrosis or cirrhosis

DISCUSSION

Acute fatty liver of pregnancy (AFLP) is a rare condition that typically occurs during the third trimester or immediately after childbirth. It is characterized by sudden clinical deterioration, presenting symptoms such as jaundice, hepatic encephalopathy, and coagulopathy, resembling acute liver failure. Women who develop AFLP often carry a heterozygous mutation in the long-chain 3-hydroxyacyl-coenzyme A dehydrogenase (LCHAD) gene.[4] LCHAD, situated on the mitochondrial membrane, plays a crucial role in the beta oxidation of long-chain fatty acids. This recessive gene mutation allows affected individuals to exhibit normal fatty acid oxidation under usual physiological conditions. However, if the foetus inherits a homozygous mutation from both parents, it is unable to efficiently oxidize fatty acids. Consequently, these fatty acids accumulate and are transferred to the mother, whose reduced enzyme function impairs their metabolism. This metabolic strain on the liver can precipitate AFLP, often alleviated by timely delivery of the infant

In the case under consideration, the patient presented critically ill. Nevertheless, swift and decisive action in intensive care led to her immediate delivery despite severe coagulopathy and other organ failures. Unfortunately, the patient experienced haemorrhagic shock due to severe coagulopathy, which was promptly addressed through re-exploration, preventing further deterioration. The noteworthy aspect of this case lies in the patient's resilience amidst multiple catastrophic events,

sustained by exemplary intensive care management and timely interventions.

Overall, this case underscores the critical role of comprehensive medical management and interdisciplinary coordination in managing AFLP and similar severe maternal conditions, thereby significantly influencing patient outcomes

CONCLUSION:

Patients suspected of having AFLP should be admitted to a facility equipped with intensive liver care capabilities, followed by expedited delivery when necessary. This approach aims to enhance survival prospects for both the mother and the baby.

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