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Acute Fatty Liver of Pregnancy Complicated by HELLP Syndrome and Disseminated Intravascular Coagulation: A Fatal Case Report

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Abstract | **Case Report**

Acute fatty liver of pregnancy (AFLP) is a rare but potentially fatal obstetric emergency that typically occurs during the third trimester or early postpartum period. It may rapidly progress to acute liver failure, severe coagulopathy, and multiorgan dysfunction. The clinical presentation often overlaps with HELLP syndrome, making diagnosis challenging. We report a fatal case of AFLP occurring in the immediate postpartum period after emergency cesarean section for HELLP syndrome. Despite aggressive intensive care management including massive transfusion and plasma exchange therapy, the patient developed refractory multiorgan failure leading to death. This case highlights the diagnostic difficulty, therapeutic limitations, and high mortality associated with fulminant AFLP.

Keywords: Acute fatty liver of pregnancy, HELLP syndrome, Liver failure, Maternal mortality, Plasma exchange, Swansea criteria.

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INTRODUCTION

Pregnancy-related liver diseases represent a small but severe group of conditions associated with significant maternal and fetal morbidity and mortality. Among these, acute fatty liver of pregnancy (AFLP) and HELLP syndrome (hemolysis, elevated liver enzymes, and low platelet count) are the most severe and life-threatening entities. AFLP is characterized by microvesicular fatty infiltration of hepatocytes leading to acute liver failure, metabolic disturbances, and coagulation abnormalities. Its pathophysiology is closely linked to defects in mitochondrial fatty acid β-oxidation, often associated with fetal fatty acid oxidation disorders.

The clinical overlap between AFLP and HELLP syndrome frequently delays diagnosis, particularly in the postpartum period, where symptoms may be attributed to obstetric complications. Early recognition and prompt multidisciplinary management are essential to improve maternal outcomes. We report a fatal case of AFLP complicating HELLP syndrome, emphasizing diagnostic challenges and management strategies in the intensive care unit.

CASE PRESENTATION

A 35-year-old multiparous woman with a history of two previous cesarean sections was admitted at 38 weeks of gestation for persistent vomiting, severe headaches, diffuse abdominal pain, and marked hypertension. The pregnancy was poorly monitored. Initial clinical and laboratory evaluation revealed severe preeclampsia complicated by HELLP syndrome, confirmed by hemolysis, elevated liver enzymes, and thrombocytopenia. An emergency cesarean section was performed due to maternal deterioration.

In the immediate postoperative period, the patient developed rapidly progressive jaundice, recurrent severe hypoglycemia, persistent abdominal pain, and neurological impairment. Laboratory investigations showed severe thrombocytopenia, prolonged prothrombin time and activated partial thromboplastin time, decreased fibrinogen levels, marked elevation of aminotransferases, mixed hyperbilirubinemia, hypoalbuminemia, and markedly elevated plasma ammonia levels.

Despite aggressive supportive treatment including transfusion of fresh frozen plasma, platelet concentrates, fibrinogen, and prothrombin complex concentrates, coagulation abnormalities persisted. Liver biopsy was contraindicated due to severe coagulopathy. Based on clinical and biological findings, the diagnosis of acute fatty liver of pregnancy was established using the Swansea criteria.

The patient underwent repeated plasma exchange sessions combined with mechanical ventilation, deep sedation, vasoactive support, and continuous metabolic correction. The clinical course was complicated by hepatic encephalopathy and progressive hemodynamic and respiratory failure. Despite maximal supportive therapy, the patient developed refractory multiorgan failure and died on day 10 of intensive care unit admission.

DISCUSSION

Acute fatty liver of pregnancy (AFLP) represents one of the most severe forms of pregnancy-associated liver disease and remains a true intensive care emergency. Although rare, AFLP is associated with rapid progression to acute liver failure, profound metabolic derangements, and multiorgan dysfunction, frequently requiring advanced organ support. Despite substantial reductions in maternal mortality over recent decades, fulminant AFLP continues to carry a high fatality rate, particularly when diagnosis is delayed or when complicated by HELLP syndrome and disseminated intravascular coagulation (DIC), as demonstrated in the present case.

Pathophysiological mechanisms: mitochondrial failure as the central driver

The pathogenesis of AFLP is now well established as a disorder of mitochondrial fatty acid β-oxidation. Pioneering work by Ibdah et al. demonstrated a strong association between AFLP and fetal fatty acid oxidation defects, most notably long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency. In this setting, the heterozygous maternal carrier state is insufficient to handle the increased lipid flux of late pregnancy, leading to accumulation of hepatotoxic fatty acid metabolites that induce diffuse microvesicular steatosis and acute hepatocellular failure.

Microvesicular steatosis impairs mitochondrial oxidative phosphorylation without significant hepatocyte necrosis, explaining the often modest transaminase elevation relative to the severity of liver dysfunction. This metabolic liver failure rapidly results in hypoglycemia, lactic acidosis, hyperammonemia, and encephalopathy. The systemic nature of mitochondrial dysfunction further explains the frequent association with renal failure, pancreatitis, and cardiopulmonary instability.

These mechanisms distinguish AFLP fundamentally from other pregnancy-related liver disorders and explain its rapid deterioration despite delivery, particularly in advanced disease.

Relationship between AFLP and HELLP syndrome: overlapping spectra

The frequent coexistence of AFLP and HELLP syndrome remains one of the most challenging aspects of diagnosis and management. Multiple large observational studies, including the UK national cohort reported by Knight et al., have demonstrated that up to one-third of AFLP cases meet diagnostic criteria for HELLP syndrome, while a subset of HELLP patients display metabolic features consistent with AFLP.

HELLP syndrome is primarily driven by placental ischemia, endothelial dysfunction, and thrombotic microangiopathy, whereas AFLP is a metabolic hepatopathy. However, both conditions converge clinically through hepatic injury, coagulopathy, thrombocytopenia, and systemic inflammation. Increasing evidence suggests that these entities may represent overlapping phenotypes within a broader spectrum of pregnancy-associated acute liver failure rather than discrete diseases.

Clinically, AFLP should be suspected when HELLP syndrome is accompanied by:

- recurrent or refractory hypoglycemia
- progressive jaundice
- marked hyperammonemia
- early encephalopathy
- disproportionate coagulopathy

The presence of these features is consistently associated with worse outcomes.

Diagnostic challenges in the critical care environment

Definitive diagnosis of AFLP remains difficult in critically ill patients. Liver biopsy, while historically diagnostic, is rarely feasible due to severe coagulopathy and thrombocytopenia. Consequently, diagnosis relies on clinical and laboratory criteria.

The Swansea criteria, first proposed and later validated by Goel et al., remain the most widely used diagnostic tool. While highly sensitive, their specificity is limited in patients with severe preeclampsia or HELLP syndrome. Nonetheless, in the ICU setting, the Swansea criteria provide a pragmatic framework to support early diagnosis when biopsy is contraindicated.

Importantly, AFLP frequently worsens in the immediate postpartum period, a phenomenon well described in multiple case series. This postpartum deterioration reflects ongoing hepatic mitochondrial failure despite removal of the placental trigger and underscores the necessity for continued ICU surveillance after delivery.

Coagulopathy and disseminated intravascular coagulation

Severe coagulopathy is a defining feature of advanced AFLP and frequently progresses to overt DIC. Impaired hepatic synthesis of coagulation factors, consumption due to systemic inflammation, and endothelial activation collectively contribute to profound hemostatic dysfunction.

Levi et al. emphasize that DIC in liver failure is complex, characterized by simultaneous bleeding and thrombotic risks. In AFLP, hypofibrinogenemia is particularly severe and strongly correlates with mortality. Conventional transfusion strategies often fail to achieve durable correction due to ongoing hepatic dysfunction, as observed in the present case.

The refractory nature of coagulopathy frequently limits invasive diagnostic and therapeutic interventions and complicates anesthetic and surgical management.

Therapeutic plasma exchange: rationale and evidence

Therapeutic plasma exchange (TPE) has emerged as an important adjunctive therapy in severe AFLP. The rationale for TPE includes removal of:
• ammonia
• bilirubin
• inflammatory cytokines
• toxic fatty acid metabolites

While simultaneously replacing deficient coagulation factors, albumin, and antithrombin.

Jin et al. reported improved survival and faster biochemical recovery in AFLP patients treated with plasma exchange compared with supportive care alone. Subsequent case series and cohort studies have reinforced these findings, particularly when TPE is initiated early in the disease course.

Evidence from acute liver failure of other etiologies further supports this approach. Larsen et al., in a randomized controlled trial, demonstrated improved transplant-free survival with high-volume plasma exchange in acute liver failure, lending biological plausibility to its use in AFLP.

Nevertheless, plasma exchange is not curative. Its effectiveness appears highly time-dependent, with limited benefit once refractory shock, advanced encephalopathy, or irreversible multiorgan failure has developed. This likely explains the poor outcome in the present case despite aggressive extracorporeal therapy.

Advanced organ support and liver transplantation considerations

While liver transplantation is rarely required in AFLP due to the potential for hepatic recovery, isolated cases have been reported in patients with irreversible liver failure. However, candidacy is often limited by multiorgan dysfunction, active coagulopathy, and hemodynamic instability.

Extracorporeal liver support systems, including molecular adsorbent recirculating systems (MARS), have been used as rescue therapy in isolated cases, though evidence remains limited. Their role remains investigational and largely confined to specialized centers.

Prognostic factors and ICU outcomes

Multiple studies have identified predictors of poor outcome in AFLP, including:
• delayed diagnosis
• hepatic encephalopathy
• renal failure
• DIC
• persistent hypoglycemia
• requirement for mechanical ventilation or vasopressors

Nelson et al. demonstrated that patients requiring renal replacement therapy or prolonged mechanical ventilation had significantly higher mortality. Despite advances in critical care, mortality in fulminant AFLP remains substantial.

Postpartum deterioration, as observed in this case, is consistently associated with adverse outcomes and highlights the importance of early ICU admission even after delivery.

Implications for intensive care practice

This case reinforces the need for heightened awareness of AFLP among intensivists. Any pregnant or postpartum patient presenting with unexplained liver dysfunction and metabolic derangements should prompt immediate consideration of AFLP.

Early diagnosis, prompt delivery, aggressive metabolic correction, early ICU admission, and timely initiation of plasma exchange or advanced liver support may improve outcomes. Given the rarity and complexity of AFLP, management in tertiary centers with multidisciplinary expertise is strongly recommended.

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