

Rapid Progression of Acute Fatty Liver of Pregnancy Ultimately Leading to Liver Transplantation: A Case Report



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Introduction

- AFLP is a rare condition (1 in 20,000 pregnancies) with high peripartum morbidity due to coagulopathy, electrolyte derangements, and concomitant multiorgan dysfunction often seen. Considering this, the labor and delivery process presents unique circumstances that create a high-risk scenario.
- Pathophysiology of AFLP focuses on fatty acid deposition, which is linked to fetal fatty acid oxidation disorders [1]. Long-chain-3-hydroxyacyl-coenzyme A dehydrogenase is responsible for β -oxidation of fatty acids. In the case of fetal deficiencies, hepatotoxic fatty acids proliferate in the fetus and cross over to maternal circulation, resulting in fat deposition in maternal hepatocytes [1]. Fetal fatty acid oxidation defects often follow autosomal recessive inheritance patterns; as such, mothers are often heterozygous, resulting in an increased susceptibility to toxic free fatty acids [1].
- Cesarean section presents an increased risk of hemorrhage but often can ensure prompt delivery.
- Meta-analyses have shown a 44% decreased maternal mortality combined with a 48% decrease in perinatal mortality in the case of C-sections [2]. Even so, C-section and vaginal deliveries have shown no associations with AFLP progression [2].
- Disease management focuses on referral to a liver transplant center and collaboration across clinical specialties to focus on the correction of coagulopathy and prompt delivery of the fetus.

Case Description

- A 32 y.o. G1P0 with dichorionic diamniotic twins presents at 33 weeks 6 days gestation with 15 hours of altered mental status with associated nausea, vomiting, and diarrhea for 2 weeks in the setting of recent IV iron transfusions.
- Labs at an outside institution were significant for INR 2.8, fibrinogen 96, total bilirubin 7.0, AST/ALT 515/347, Cr 2.26, lactate dehydrogenase 637, and lactate 4.0. She was immediately transferred to a tertiary care center for escalation of care.
- Upon arrival, she was immediately evaluated by a multidisciplinary including OBGYN, Critical Care, and Anesthesia teams.
- The decision was made to induce labor in this patient given the patient’s coagulopathy. AFLP was of concern at this time.
- Neuraxial anesthesia was not offered to the patient secondary to coagulopathy. She was placed on a PCA. As she progressed towards delivery she became increasingly encephalopathic and coagulopathic. She was transfused multiple units of both cryoprecipitate and FFP in an attempt to correct her coagulopathy.
- Patient was transferred to OR for second stage of labor in anticipation of vaginal delivery.
- Baby A was successfully delivered vaginally. However, Baby B due to the patient’s poor effort in the setting of increasing encephalopathy patient had a prolonged second stage.
- Due to maternal encephalopathy, the OBGYN team decided to transfer the patient to the ICU in anticipation of Baby B delivery.
- Due to abnormal coagulation status OBGYN permitted prolonged second stage of labor for Baby B (4 hours).
- She was then immediately transferred to a transplant center where she would ultimately undergo liver transplantation 2 days after delivery in the setting of acute fatty liver of pregnancy (AFLP).

Hepatic Condition	History of Liver Disease	Hypertension	Transaminitis	Elevated INR
AFLP	No	Maybe	Yes	Yes
Viral Hepatitis	Yes	No	Yes	Yes
HELLP Syndrome	No	Yes	Maybe	Maybe
Intrahepatic Cholestasis of Pregnancy	No	No	Maybe	No
Gastroenteritis	No	No	Maybe	No

Figure 1: Features of hepatic conditions that may effect pregnancy. AFLP: Acute fatty liver of pregnancy; HELLP: hemolysis, elevated serum level of enzymes, and low platelets syndrome

Anesthetic Planning & Management

- Neuraxial anesthesia is the method of choice in most obstetrical patients as it is an extremely effective means of providing pain relief with minimal risk factors to both the mother and baby.
- Neuraxial anesthesia has also shown to reduce the risk of postpartum hemorrhage, which clearly would have benefited this patient [3].
- In this case the patient’s disease status (coagulopathy) precluded the use of neuraxial anesthesia as the patient would have been at high risk for complications. As a result the anesthetic management of this patient focused on general endotracheal anesthesia if a cesarean delivery were needed.
- Blood products were immediately made available with the goal to imminently deliver both fetuses while stabilizing the patient for transfer to a liver transplant center. As described previously, the patient did not ultimately hemorrhage peripartum nor required a cesarean delivery.

Discussion

- This case presents specific challenges.
 - The patient’s multiple gestations create particularly increased risks.
 - The patient was transferred to a tertiary center without liver transplantation immediately available.
 - The goal at this center was to stabilize the patient’s coagulopathy while the patient progressed toward delivery to transfer her for transplant evaluation.
 - The OB team prioritized patient vaginally delivery to minimize the risk of postpartum hemorrhage given her profound coagulopathies.
- With regard to obstetrical management, these patients often necessitate prompt delivery given the progression of AFLP.
- This patient ultimately had increasing encephalopathy which led to poor effort. This may have contributed to delays in delivery, which is associated with fetal and maternal mortality [1].
- Even so, coagulopathy is of great concern in these patients; thus making a cesarean delivery inherently undesirable as well.
- Literature review revealed that 65% of AFLP cases undergo cesarean section [1].
 - It is critical that's patient’s individualized delivery plan is individualized as many factors must be considered prior to deciding delivery mode (i.e. like the acuity of the situation, maternal disease progression, fetal status, and provider experience).
- AFLP presents particular challenges that can create a dire situation for parturients and fetal outcomes. Coagulopathy and encephalopathy are challenges that make anesthetic management difficult. In these cases, anesthesiologists must be involved in perioperative planning and be immediately available given the expertise possessed in resuscitation, airway management, and obstetrical care.

References

