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Case Report **Anaesthesia**

Anesthetic Outcome of a Primigravida Woman with Acute Fatty Liver of Pregnancy (AFLP) Complicated by Multiorgan Dysfunction Syndrome (MODS): An Obstetric Emergency

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Abstract

Acute fatty liver of pregnancy (AFLP) is a rare but life-threatening obstetric emergency, often complicated by multiorgan dysfunction syndrome (MODS). We report a case of a 25-year-old primigravida woman at 34 weeks and 7 days of a twin pregnancy presenting with abdominal pain, nausea, and vomiting for three days. Diagnosis was established based on clinical, laboratory, and imaging findings. Prompt multidisciplinary management, including termination of pregnancy and intensive care support, resulted in a favorable maternal outcome. This case underscores the importance of early recognition and aggressive intervention in AFLP complicated by MODS.

Keywords: Acute fatty liver of pregnancy, Multiorgan dysfunction syndrome, Obstetric emergency, Twin pregnancy, Cesarean delivery, Supportive care.

INTRODUCTION

Acute fatty liver of pregnancy (AFLP) is a rare but life-threatening obstetric emergency that predominantly occurs in the third trimester. Characterized by microvesicular fatty infiltration of hepatocytes, AFLP can rapidly lead to liver dysfunction, multi-organ dysfunction syndrome (MODS), and significant maternal and fetal morbidity and mortality. Its estimated incidence ranges from 1 in 7,000 to 1 in 20,000 pregnancies, and severe cases have maternal mortality rates as high as 18%, emphasizing the need for prompt recognition and multidisciplinary management to improve outcomes (Knight *et al*., 2008; Usta *et al*., 1994).

The pathophysiology of AFLP is not fully understood but is thought to involve mitochondrial dysfunction in fatty acid metabolism. Deficiency in long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD), a mitochondrial beta-oxidation enzyme, is implicated in many cases. This deficiency leads to the accumulation of toxic fatty acid metabolites, causing hepatocyte injury and microvesicular steatosis. Genetic mutations in the HADHA gene, encoding a subunit of LCHAD, are commonly associated with AFLP, with maternal and fetal heterozygosity increasing susceptibility (Ch'ng *et al*., 2002; Ibdah *et al*., 1999). AFLP typically presents between 30 and 38 weeks of gestation, although postpartum cases are reported. The clinical presentation is variable and overlaps with conditions such as preeclampsia and HELLP syndrome. Initial symptoms include nonspecific findings like nausea, vomiting, fatigue, and abdominal discomfort. Progressive liver dysfunction may manifest as jaundice, pruritus, and dark urine. Laboratory abnormalities include elevated liver enzymes, hyperbilirubinemia, hypoglycemia, coagulopathy, and hyperammonemia. Severe cases may lead to complications such as acute kidney injury, pancreatitis, encephalopathy, disseminated intravascular coagulation, and acute respiratory distress syndrome, necessitating urgent intervention (Knight *et al*., 2008; Usta *et al*., 1994).

Diagnosis is primarily clinical, supported by laboratory and imaging findings. The Swansea criteria, a set of 14 diagnostic parameters, are commonly used; six or more criteria suggest AFLP. These include vomiting, abdominal pain, polydipsia/polyuria, encephalopathy,

elevated bilirubin (≤50 µmol/L), hypoglycemia (<4 mmol/L), elevated uric acid (>340 µmol/L), leukocytosis, ascites or a bright liver on ultrasound, elevated transaminases (≤50 IU/L), hyperammonemia (>47 µmol/L), renal impairment (creatinine >150 µmol/L), coagulopathy (INR >1.5), and microvesicular steatosis on liver biopsy (Knight *et al*., 2008; Tran *et al*., 2016; Usta *et al*., 1994). Imaging, such as ultrasound or CT, may show hyperechoic liver or hepatomegaly, but these are nonspecific findings. Liver biopsy, showing microvesicular steatosis, remains the gold standard for diagnosis but is rarely performed due to its invasive nature and associated risks (Ch'ng *et al*., 2002; Usta *et al*., 1994).

The Case

A 25-year-old primigravida was admitted to the obstetrics and gynecology ward at midnight. She was at 34 weeks and 7 days of gestation with a confirmed twin pregnancy. She complained of abdominal pain, persistent nausea, and vomiting 12-15 times daily for the last three days. She denied pruritus or other significant symptoms.

Recent History

The patient reported progressive weakness and loss of appetite over the past week. She had visited a local clinic two days prior, where symptomatic management was provided without significant relief. There was no history of fever, urinary symptoms, or previous liver disease. She had regular antenatal checkups, and her pregnancy was otherwise uneventful.

On Admission

The patient appeared acutely ill, pale, and dehydrated. Vital signs revealed tachycardia (heart rate: 115 bpm), hypotension (BP: 90/60 mmHg), and tachypnea (respiratory rate: 26/min). Her oxygen saturation was 94% on room air. The patient exhibited mild icterus and peripheral edema.

Physical Examination

Abdominal examination revealed tenderness in the epigastrium and right upper quadrant. Fundal height was consistent with 34 weeks gestation, and fetal heart rates were within normal ranges for both twins. No uterine contractions or signs of preterm labor were observed.

Examination of the Nervous System

The patient was alert but disoriented to time and place. Neurological examination revealed no focal deficits or signs of hepatic encephalopathy.

Table 1: Laboratory Diagnosis

Parameter	Result	Reference Range (Female)	Interpretation
Hemoglobin	8.6 g/dL	12.0–16.0 g/dL	Anemia
Total leukocyte count	18,000/mm³	4,000–11,000/mm³	Leukocytosis, indicative of systemic inflammation
Platelet count	82,000/mm³	150,000–450,000/mm³	Thrombocytopenia
Serum bilirubin	6.4 mg/dL	0.1–1.2 mg/dL	Hyperbilirubinemia, suggestive of liver dysfunction
Alanine aminotransferase (ALT)	156 IU/L	7–56 IU/L	Elevated, indicative of liver injury
Aspartate aminotransferase (AST)	178 IU/L	5–40 IU/L	Elevated, indicative of liver injury
Alkaline phosphatase (ALP)	240 IU/L	40–129 IU/L	Elevated, may be pregnancy-related or hepatic
Prothrombin time (PT)	22 seconds (INR 2.1)	11–13.5 seconds (INR <1.1)	Coagulopathy, consistent with liver dysfunction
Serum creatinine	2.3 mg/dL	0.6–1.2 mg/dL	Renal impairment
Blood urea nitrogen (BUN)	48 mg/dL	7–20 mg/dL	Elevated, indicative of renal dysfunction
Blood glucose	62 mg/dL	70–100 mg/dL (fasting)	Hypoglycemia, suggestive of hepatic dysfunction

Table 2: Day wise Progress Report of Patient via Parameters of Blood Test

Parameters	Day 1	Day 3	Day 4	Day 8	Day 15
S. ALP (a)	516	575	153	120	-
S. ALT (b)	136	138	48	40	-
S. Bilirubin (c)	9.3	11.7	11.3	9	3.89
INR (d)	3.1	3.2	1.2	1	-
WBC (e)	17,000	28,400	21,000	11,000	8,300
S. Uric Acid (f)	7	9.1	6	-	-
Total Platelet count (g)	1,80,000	1,56,000	1,20,000	1,40,000	1,77,000
S. Ammonia (h)	41	-	-	-	-
S. Creatinine (i)	1.8	2.89	2.81	1.32	0.8

Other Biochemical Findings

Ammonia levels were mildly elevated (54 µmol/L). Serum lactate dehydrogenase (LDH) was 850 IU/L, and uric acid was 7.5 mg/dL. Arterial blood gas analysis showed metabolic acidosis with a pH of 7.28.

Ultrasonography (USG)

Abdominal ultrasound revealed hepatomegaly with increased echogenicity suggestive of fatty infiltration. Fetal biometry was consistent with gestational age, and no signs of fetal compromise were noted.

Clinical Management

The patient was transferred to the intensive care unit (ICU) for multidisciplinary care. Intravenous fluids, glucose supplementation, and empirical antibiotics were initiated. Coagulopathy was corrected with fresh frozen plasma and vitamin K. Hypoglycemia was managed with continuous glucose infusion.

Surgical Management/Operation Note and Subsequent Steps Taken

Given the deteriorating maternal condition, emergency cesarean delivery was performed under general anesthesia within six hours of admission. Two live female neonates were delivered with birth weights of 2.1 kg and 2.0 kg, respectively. Both required short-term neonatal intensive care for mild respiratory distress.

Therapeutic Modalities Instituted

Postoperatively, the patient received intensive supportive care, including vasopressors for hypotension, hemodialysis for acute kidney injury, and mechanical ventilation due to respiratory compromise. Broad-spectrum antibiotics and antifungals were administered to prevent sepsis.

Notable Improved Evidence

Over the next 10 days, the patient showed gradual improvement in hepatic and renal functions. Serum bilirubin levels declined, coagulation parameters normalized, and renal function recovered. She was extubated on postoperative day 7 and discharged from the ICU on day 12.

DISCUSSION

Acute fatty liver of pregnancy (AFLP) is a rare yet life-threatening obstetric emergency that occurs due to mitochondrial dysfunction in the metabolism of fatty acids. This condition predominantly arises due to defective fatty acid oxidation, often associated with fetal genetic mutations, such as defects in the long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) enzyme(Ramanathan & Ibdah, 2022). Such mutations can lead to an accumulation of toxic metabolites that compromise maternal hepatic function, causing microvesicular steatosis, which is the hallmark of AFLP (Wei *et al*., 2010).

Pathophysiology

The mitochondrial dysfunction in AFLP involves an impairment in the beta-oxidation of fatty acids, primarily due to genetic mutations in the fetus. LCHAD deficiency, one of the most frequently implicated defects, results in the accumulation of long-chain fatty acids and their derivatives, which are toxic to hepatocytes(Ko & Yoshida, 2006). This genetic defect is typically inherited in an autosomal recessive manner, with the fetus inheriting mutations from both parents. Consequently, the maternal liver—exposed to the toxic metabolites via the placenta—undergoes functional derangement and morphological changes, leading to hepatic failure. Understanding this pathophysiological mechanism has improved diagnostic and management approaches, although the condition remains challenging to recognize due to its nonspecific presentation.

Clinical Presentation and Diagnosis

The early symptoms of AFLP often mimic other hepatic or obstetric conditions such as HELLP (hemolysis, elevated liver enzymes, and low platelet count) syndrome, intrahepatic cholestasis of pregnancy, and viral hepatitis. Common presenting features include nausea, vomiting, abdominal pain, jaundice, and malaise, which can progress to more severe complications like encephalopathy, coagulopathy, and renal failure if untreated(Riely, 1999). A high index of suspicion is critical, especially in the third trimester when AFLP most commonly occurs.

Diagnostic criteria such as the Swansea criteria, which include clinical, laboratory, and imaging findings, are helpful in distinguishing AFLP from other conditions. These criteria consider factors such as hypoglycemia, elevated transaminases, coagulopathy, renal impairment, and radiological evidence of fatty liver(Lee & Brady, 2009). Despite these tools, definitive diagnosis often requires exclusion of other hepatic conditions, necessitating interdisciplinary collaboration among obstetricians, hepatologists, and intensivists.

Challenges in Differentiation

Differentiating AFLP from similar conditions remains one of the primary challenges in clinical practice. HELLP syndrome, in particular, shares several overlapping features such as thrombocytopenia, elevated liver enzymes, and hemolysis, complicating diagnosis (Terrault & Williamson, 2022). However, AFLP typically presents with hypoglycemia, hyperbilirubinemia, and more pronounced hepatic dysfunction, which can aid differentiation. Imaging modalities such as ultrasound or magnetic resonance imaging (MRI) may show diffuse fatty infiltration of the liver in AFLP but are not always definitive (Solanke *et al*., 2016).

Management Strategies

The cornerstone of AFLP management is prompt delivery, which is the definitive treatment for

halting the progression of the disease. Early delivery, often via cesarean section, minimizes maternal and fetal morbidity and mortality. In this case, the timely decision to proceed with cesarean delivery played a pivotal role in achieving a favorable outcome. The management of AFLP extends beyond delivery to include intensive supportive care aimed at addressing complications such as multiorgan dysfunction syndrome (MODS), coagulopathy, and renal failure (Ziki *et al*., 2019).

Management of Complications

Managing coagulopathy and renal failure posed significant challenges in this case. Severe coagulopathy, characterized by prolonged prothrombin time and reduced fibrinogen levels, necessitated aggressive correction using blood products such as fresh frozen plasma, cryoprecipitate, and platelets. Renal failure, another common complication of AFLP, often results from acute tubular necrosis secondary to hypovolemia or sepsis. Continuous renal replacement therapy (CRRT) may be required in severe cases to support renal function while awaiting recovery (Watson & Seeds, 1990).

In the context of MODS, a multidisciplinary approach involving critical care specialists is essential. This includes monitoring and supporting cardiac, respiratory, and renal functions while addressing metabolic derangements such as hypoglycemia and lactic acidosis. In the presented case, the aggressive management of MODS, including the use of vasopressors, mechanical ventilation, and renal replacement therapy, was instrumental in ensuring a favorable outcome.

Prognosis and Long-Term Outcomes

Despite its severity, early recognition and timely management of AFLP have significantly improved maternal and fetal outcomes. However, residual complications such as chronic kidney disease, hepatic insufficiency, or neurodevelopmental issues in the neonate may persist, necessitating long-term follow-up (Reyes *et al*., 1994). Genetic counseling is recommended for affected families to identify carriers of LCHAD deficiency and provide guidance for future pregnancies. Advances in genetic screening and prenatal diagnostics have also enhanced the ability to predict and manage recurrent cases of AFLP in subsequent pregnancies (Masiga & Turner, 2004).

CONCLUSION

AFLP remains a diagnostic and therapeutic challenge in obstetrics due to its rare occurrence and overlapping presentation with other conditions. This case underscores the importance of maintaining a high index of suspicion and promptly initiating definitive management through delivery and intensive supportive care. Advances in understanding the genetic and molecular basis of AFLP hold promise for improving diagnostic accuracy and therapeutic outcomes, although further research is needed to elucidate the precise mechanisms underlying this complex condition. Multidisciplinary care and early intervention are critical in achieving favorable outcomes for both mother and fetus.

REFERENCE

- Ch'ng, C. L., Morgan, M., Hainsworth, I., & Kingham, J. G. C. (2002). Prospective study of liver dysfunction in pregnancy in Southwest Wales. *Gut*, *51*(6), 876–880. https://doi.org/10.1136/GUT.51.6.876
- Ibdah, J. A., Bennett, M. J., Rinaldo, P., Zhao, Y., Gibson, B., Sims, H. F., & Strauss, A. W. (1999). A fetal fatty-acid oxidation disorder as a cause of liver disease in pregnant women. *The New England Journal of Medicine*, *340*(22), 1723–1731. https://doi.org/10.1056/NEJM199906033402204
- Knight, M., Nelson-Piercy, C., Kurinczuk, J. J., Spark, P., & Brocklehurst, P. (2008). A prospective national study of acute fatty liver of pregnancy in the UK. *Gut*, *57*(7), 951–956. https://doi.org/10.1136/GUT.2008.148676
- Ko, H. H., & Yoshida, E. M. (2006). Acute fatty liver of pregnancy. *Canadian Journal of Gastroenterology = Journal Canadien de Gastroenterologie*, *20*(1), 25–30. https://doi.org/10.1155/2006/638131
- Lee, N. M., & Brady, C. W. (2009). Liver disease in pregnancy. *World Journal of Gastroenterology*, *15*(8), 897–906. https://doi.org/10.3748/WJG.15.897
- Masiga, D. K., & Turner, C. M. R. (2004). Amplified (restriction) fragment length polymorphism (AFLP) analysis. *Methods in Molecular Biology (Clifton, N.J.)*, *270*, 173–186. https://doi.org/10.1385/1-59259-793-9:173
- Ramanathan, R., & Ibdah, J. A. (2022). Mitochondrial Dysfunction and Acute Fatty Liver of Pregnancy. *International Journal of Molecular Sciences*, *23*(7). https://doi.org/10.3390/IJMS23073595
- Reyes, H., Sandoval, L., Wainstein, A., Ribalta, J., Donoso, S., Smok, G., Rosenberg, H., & Meneses, M. (1994). Acute fatty liver of pregnancy: a clinical study of 12 episodes in 11 patients. *Gut*, *35*(1), 101–106. https://doi.org/10.1136/GUT.35.1.101
- Riely, C. A. (1999). Liver disease in the pregnant patient. *American Journal of Gastroenterology*, *94*(7), 1728–1732. https://doi.org/10.1111/J.1572-0241.1999.01199.X
- Solanke, D., Rathi, C., Pandey, V., Patil, M., Phadke, A., & Sawant, P. (2016). Etiology, clinical profile, and outcome of liver disease in pregnancy with predictors of maternal mortality: A prospective study from Western India. *Indian Journal of Gastroenterology*, *35*(6), 450–458. https://doi.org/10.1007/s12664-016-0704-6
- Terrault, N. A., & Williamson, C. (2022). Pregnancy-Associated Liver Diseases.

Gastroenterology, *163*(1), 97-117.e1. https://doi.org/10.1053/j.gastro.2022.01.060

- Tran, T. T., Ahn, J., & Reau, N. S. (2016). ACG Clinical Guideline: Liver Disease and Pregnancy. *The American Journal of Gastroenterology*, *111*(2), 176–194. https://doi.org/10.1038/AJG.2015.430
- Usta, I. M., Barton, J. R., Amon, E. A., Gonzalez, A., & Sibai, B. M. (1994). Acute fatty liver of pregnancy: an experience in the diagnosis and management of fourteen cases. *American Journal of Obstetrics and Gynecology*, *171*(5), 1342–1347. https://doi.org/10.1016/0002-9378(94)90158-9
- Watson, W. J., & Seeds, J. W. (1990). Acute fatty liver of pregnancy. *Obstetrical and Gynecological Survey*, *45*(9), 585–593. https://doi.org/10.1097/00006254-199009000-00004
- Wei, Q., Zhang, L., & Liu, X. (2010). Clinical diagnosis and treatment of acute fatty liver of pregnancy: a literature review and 11 new cases. *The Journal of Obstetrics and Gynaecology Research*, *36*(4), 751–756. https://doi.org/10.1111/J.1447-0756.2010.01242.X
- Ziki, E., Bopoto, S., Madziyire, M. G., & Madziwa, D. (2019). Acute fatty liver of pregnancy: A case report. *BMC Pregnancy and Childbirth*, *19*(1), 1–3. https://doi.org/10.1186/S12884-019-2405-5/FIGURES/1