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CASE REPORT

A SEVERE CASE OF ACUTE FATTY LIVER OF PREGNANCY COMPLICATED BY POSTPARTUM PREECLAMPSIA AND SEPSIS

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ABSTRACT

Acute fatty liver of pregnancy is an uncommon life threatening obstetric emergency associated with significant morbidity and mortality. We report a case of a 20-year-old pregnant lady who presented with nausea, vomiting, epigastric pain, and jaundice. A clinical diagnosis of AFLP was made and supportive management was provided. This case highlights the need for disease recognition and treatment of acute fatty liver of pregnancy, and the clinical and biochemical overlap between AFLP and the syndrome of hemolysis, elevated liver enzymes, and low platelet count in pregnancy.

INTRODUCTION

Acute fatty liver of pregnancy (AFLP) is an uncommon life threatening obstetric emergency with an estimated incidence of 1 in 7000 to 1 in 20,000 live births (1).Sheehan first described the disease in 1940 (2).AFLP is characterized by sudden onset of liver failure in the third trimester or the post-partum period. The exact etiology is unknown but defects in β-oxidation of fatty acids may be responsible (3). Twin pregnancies, body mass index <20, older age, and male fetus are possible risk factors (4). Preeclampsia is an accompanying diagnosis in 21-64% of cases (4, 5). Women with AFLP commonly present with nausea, vomiting, abdominal pain, and jaundice(4-9). Laboratory testing reveals elevations in bilirubin, aminotransferase, uric acid, and creatinine, and the presence of coagulopathy, thrombocytopenia, and hypoglycemia (6-12). Severe cases of AFLP manifest with multiorgan failure including acute respiratory distress syndrome (ARDS), hepatic failure, renal failure sepsis, and ongoing coagulopathy(13,14).Early diagnosis and prompt treatment including delivery can lessen morbidity and mortality (10, 15).
We present a severe case of AFLP complicated by postpartum preeclampsia and sepsis.

CASE SUMMARY

A 20 year-old primigravida with a twin pregnancy (gestational ages 34 weeks) presented to a regional hospital in labor. One week prior to admission, she experienced vomiting, epigastric pain, and blurred vision; two days prior to presentation, she developed jaundice.

The patient underwent an emergency Caesarean section because of cord prolapse; two low birth weight babies were delivered but one died after a few hours. Her post-operative course was complicated by post-partum preeclampsia: blood pressure (BP) of 145/95 and 4+ urine protein), incisional bleeding, and altered mental status. She was treated with IV magnesium and transferred to TikurAnbessa Specialized Hospital (TASH) for further care.

Upon arrival at the TASH Emergency Department and subsequent rapid transfer to the Obstetric Ward, the patient was acutely ill-appearing and confused; her BP was 100/70, pulse rate 110/minute, respiratory rate 16/minute, T 36.5^0C, and oxygen saturation (room air) 99%. Over the next several hours, she became more tachypnic, tachycardic and hypoxemic. She was transferred to Medical Intensive Care Unit (MICU).

In the MICU, her BP was 100/60 mm Hg; she was confused, jaundiced with pale conjunctiva, and had a large hematoma at the incision site. Given risk factors of primipara with multiple gestations, her clinical presentation, and laboratory data (Table 1), a presumptive diagnosis of pre-eclampsia with hemolytic anemia, elevated liver enzymes and low platelets (HELLP) and/or AFLP was made.
Over the next 4 days, her condition worsened. She was intubated and mechanically ventilated for respiratory failure and aspiration pneumonia. A diagnosis of sepsis was made after her BP dropped to 80/50 mm Hg despite normal saline administration; she continued to exhibit confusion and had a rising creatinine with a decreased urine output (300 ml in 24 hours).

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Blood cultures grew Acinetobacter and she was started on Meropenem. Hemodialysis was begun for acute renal failure (creatinine 5.6 mg/dL).Her mental status deteriorated further and she was treated with lactulose and Metronidazole for possible clinical hepatic encephalopathy (ammonia level was not available). She continued to have marked elevation of her bilirubin and aminotransferases (maximum values: total bilirubin-24.4 mg/dl; AST-226IU/L, and ALT-174IU/L), and hypoalbuminemia (2.3 g/dL).

Her platelet count dropped to 12 x 10^9/L and her incisional hematoma enlarged, requiring evacuation. She received several liters of saline, 6 units of packed red blood cells (PRBC), and 12 units of fresh frozen plasma (FFP). Abdominal ultrasound was normal.

The laboratory findings on admission are summarized below (Table 1):

Table 1: Admission laboratory results

Value	Result	Reference values
Hemoglobin	8.7 g/dL	12-15 g/Dl
WBC	31,000/mm3	4,000-10,000/mm3
Platelets	100 x 109/L	150-450 x109/L
PT	59.3 sec	11-14 sec
aPTT	150 sec	25-35 sec
INR	5.35	<=1.1
Total Bilirubin	11.8 mg/dL	0.2-0.8 mg/dL
Indirect Bilibubin	3.9 mg/dL	0.0-0.3 mg/dl
Albumin	2.3 g/dL	3.5 to 5.5 g/dL
AST	151 IU/L	8-40 IU/L
ALT	97 IU/L	7-56 IU/L
LDH	1560 mg/dL	100-190 mg/dL
Creatinine	4.2 mg/dL	0.4 – 1.1 mg/dL
Uric acid	12.6 mg/dL	3.4-7 mg/dL
Random Blood Sugar	61 mg/dL	70-100 mg/dL
HBsAg	Negative	
Anti HCV Antibody	Negative	
Anti HAV Antibody	Negative	
Anti HEV Antibody	Negative	

Remarkably, over the next 21 days, the patient gradually improved with supportive care. She was extubated on hospital day 25. Shortly thereafter, she was well enough to be discharged home. Laboratories at discharge are shown in Table 2. At her six-week outpatient follow-up visit, she had normal liver and renal function studies.

DISCUSSION

The diagnosis of AFLP is usually made clinically and can be based on six or more of the following Swansea criteria: vomiting; abdominal pain; polydipsia/polyuria; encephalopathy; elevated bilirubin >0.8 mg/dL; hypoglycemia <72 mg/dL; elevated uric acid>5.7 mg/dL; leukocytosis >11x10^9/L; ascites or bright liver on ultrasound; elevated transaminases (> 42 IU/L); elevated ammonia >47 μmol/L; renal impairment creatinine>1.7 mg/dL; coagulopathy (PT >14 sec or APTT >34 sec), or microvesicularsteatosis on liver biopsy(8, 16).

Our patient met 10 of these criteria. Furthermore, our patient had manifestations of severe AFLP with multiorgan failure including respiratory failure requiring mechanical ventilation, sepsis, hepatic encephalopathy, renal failure requiring dialysis, and coagulopathy as seen in other similar severe cases of AFLP(13,14).

Ultrasound and CT scans of the liver have been recommended in the workup of AFLP, but the specificity and sensitivity of these studies are insufficient to make a diagnosis, and the likelihood of false negative results is high (4). Our patient had a normal abdominal ultrasound. Liver biopsy can be diagnostic showing micro vesicular fatty infiltration of the hepatocytes; however the procedure is usually unnecessary and can be ill advised in the setting of a coagulopathy or thrombocytopenia (9, 15,17).

There are several other liver disorders of pregnancy that need to be considered in the differential diagnosis. HELLP syndrome presents similarly with elevated liver enzymes and a low platelet count in the third trimester. However with HELLP, hemolysis is often seen. Our patient did not have evidence of hemolysis on peripheral blood smear and her indirect bilirubin was relatively low. Hypoglycemia, encephalopathy, and coagulopathy, seen in our patient, are more common with AFLP. Other more common liver disorders of pregnancy include hyperemesis in the first trimester, cholestasis of pregnancy in the second trimester, and viral or drug induced hepatitis, gallstone disease, or malignancy in any trimester. Our patient did not have features of any of these disorders.

Additionally, there was no evidence of ischemic hepatitis; our patient developed jaundice prior to requiring PRBC transfusions and she never required catecholamine support for hypotension. Treatment of AFLP is focused on maternal stabilization and delivery of the fetus, regardless of the gestational age. With this approach, maternal and neonatal mortality rates have markedly decreased. The maternal mortality rate due to this disease has decreased from 80-85% to 7-18% and the fetal morality rate from 50% to 9-23% (16,18,19).

Conclusion: Our patient presented with a severe case of AFLP complicated by postpartum preeclampsia and sepsis.

She experienced a full recovery after prompt delivery by Cesarean section, which was performed for cord prolapse and ongoing maternal support. This case highlights the need for disease recognition and treatment of AFLP, and the clinical and biochemical overlap between AFLP and HELLP syndrome.

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