

Acute Fatty Liver of Pregnancy: A Confusing Clinical Entity

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Acute fatty liver of pregnancy (AFLP) is a rare disease that is unique to pregnancy. AFLP occurs late in pregnancy and is characterized by signs of acute hepatic failure with non-specific symptoms such as nausea, vomiting, fatigue, thirst, headache, and altered mental status. If untreated, AFLP can lead to fulminant hepatic failure, coagulopathy, hemorrhage, and death. This article discusses a case of AFLP in a 29-year-old woman. A discussion of etiology, pathophysiology, diagnosis, and treatment is also presented.

CASE PRESENTATION

A 29-year-old primigravida, para 0 woman at 31 weeks gestation presents to her obstetrician complaining of progressive malaise, myalgia, severe fatigue, nausea, and vomiting. The patient is also experiencing right upper quadrant pain and her urine is dark brown in color.

The patient has experienced no complications with her pregnancy. Her medical history is not significant. Social history is marked by frequent consumption of shrimp and tuna. The patient is currently taking calcium carbonate for nausea.

Physical Examination

The patient is referred to an infectious disease specialist for possible hepatitis evaluation. On physical examination, the patient appears ill and is unable to stand without assistance. She is afebrile with orthostatic changes on vital signs. The patient is positive for scleral icterus, mild right upper quadrant tenderness, and normal fundal height (31 cm).

Hospital Admission and Laboratory Evaluation

The patient is admitted to the hospital. Fetal death in utero is diagnosed. Laboratory examination of the patient reveals the following values:

- Leukocyte count, 20,800/mm³ with 6% band forms, 14% lymphocytes, and 4% monocytes
- Hemoglobin, 12.7 g/dL
- Hematocrit, 37.1%
- Platelets, 187,000/mm³
- Alanine aminotransferase, 523 U/L
- Aspartate aminotransferase, 688 U/L
- Total bilirubin, 11.5 mg/dL
- Phosphatase (alkaline), 432 U/L
- Lactate dehydrogenase, 466 U/L
- Gamma glutamyltransferase, 348 U/L
- Prothrombin time, 20.3 seconds
- Partial thromboplastin, 68.7 seconds
- Fibrinogen, 23 mg/dL
- Sodium, 125 mEq/L
- Potassium, 3.3 mEq/L
- Chloride, 86 mEq/L
- Bicarbonate, 30 mEq/L
- Blood urea nitrogen, 10 mg/dL
- Creatinine, 1.4 mg/dL
- Calcium, 11.3 mg/dL
- Glucose, 43 mg/dL

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- Triglycerides, 164 mg/dL
- Cholesterol, 226 mg/dL
- Uric acid, 5.8 mg/dL

Screening is negative for TORCH (toxoplasmosis, other [syphilis, hepatitis, zoster], rubella, cytomegalovirus, and herpes simplex [maternal infections]) titers, hepatitis A virus, IgM and IgG, hepatitis B surface antigen, hepatitis B core antibody, hepatitis B surface antibody, and hepatitis C virus. No gallstones are detected on ultrasonography. The diagnosis of AFLP is made based on the patient's profound hypoglycemia, moderate elevation of the results of liver function tests, and normotensive state.

Treatment

Oxytocin is given to induce labor, and a 3-lb, 11-oz male fetus is delivered. The patient's condition steadily improves after the delivery. On hospital day nine the patient is discharged home after general improvement in laboratory results.

DISCUSSION

Stander and Cadden¹ described the first case of AFLP in 1934. In 1996, Castro et al² estimated the incidence of AFLP to be one in 6692 deliveries over a 12-year period. Although rare, AFLP is a life-threatening complication of pregnancy with a maternal mortality rate of almost 20% and a fetal mortality rate of 23%.^{1,3}

AFLP typically occurs in primigravidas in their third trimester between weeks 36 and 40. AFLP is extremely similar to preeclampsia and may even be the same disease on a broader continuum,⁴ which potentially explains the increased incidence of disease in woman in the third trimester.

Etiology

The exact etiology of AFLP is unknown, but evidence points to hepatic damage caused by defects in the activity of long chain 3-hydroxyacyl coenzyme A dehydrogenase (LCHAD). Toxins produced from aberrant fatty acid oxidation may play a role in the development of AFLP. Moreover, the interaction between an LCHAD activity-deficient female and similarly deficient fetus may also lead to the progression to AFLP and could explain the rarity of the disease.^{5,6}

Pathophysiology and Patient Presentation

The pathophysiology of AFLP is related to the coagulopathy common in other types of acute liver failure. Patients typically present with unremitting nausea, vomiting, weight loss, and abdominal pain. Jaundice usually

develops after several days.^{1,3} Abnormal laboratory findings include moderate elevations of aminotransferase levels (300 to 500 U/L), bilirubin ranging from 3 to 25 mg/dL, markedly elevated alkaline phosphatase levels (normal, 42 to 98 U/L), and leukocytosis.^{1,2} Abnormal coagulation parameters are found in association with secondary disseminated intravascular coagulation. Profound hypoglycemia occurs as liver dysfunction progresses.³

Diagnosis

Previously, liver biopsy was required for definitive diagnosis of AFLP because of the disease's indistinguishable clinical similarity to fulminant viral hepatitis; however, serologic determinants of viral hepatitis have made liver biopsy unessential.

Treatment

Delivery is the definitive treatment for AFLP. Induction may suffice, but cesarean section may be required. Patients with AFLP generally improve soon after delivery unless hepatic encephalopathy has developed.

SUMMARY

In cases of AFLP, a good maternal outcome is dependent on early diagnosis and immediate delivery. The patient in this case study survived, however, the fetus died. The outcome was likely a result of the disruption of metabolic homeostasis as related to, but not inclusive of, severe hypoglycemia. Although AFLP is rare, physicians must consider this diagnosis in any patient presenting with symptoms resembling acute viral hepatitis during the third trimester. HP

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