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Original Research Paper

A Case of Fulminant Acute Fatty Liver of Pregnancy

MEDICAL SCIENCE

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ABSTRACT

Acute fatty liver of pregnancy(AFLP) is a serious complication of late third trimester of pregnancy, characterized by microvesicular steatosis of liver leading to acute liver insufficiency. It is more common in nulliparous with a male fetus. It generally occurs in women heterozygous for Long-Chain 3-hydroxy acyl-coenzyme A dehydrogenase (LCHAD), which is necessary for of fatty acid oxidation. Patients generally presents with non specific complaints of malaise, nausea, vomiting, right hypochondriac pain, jaundice. In severe cases, multisystem involvement including acute renal failure, encephalopathy, gastrointestinal bleeding, pancreatitis, coagulopathy, hypoglycemia occurs. The most striking feature is high level of bilirubin associated with moderate increase of transaminases. Early diagnosis, prompt delivery and intensive supportive care are the cornerstones in the management of AFLP. Maternal mortality rates in the past were 75% but with prompt diagnosis and timely treatment mortality rate has decreased to approximately 18%. Neonatal mortality rates are usually higher (60 to 85%).

Introduction:

Acute fatty liver of pregnancy (AFLP) first described by Sheehan in 1940, is a serious complication unique to human pregnancy characterized by microvesicular steatosis of liver without inflammation and necrosis leading to acute liver insufficiency. It is a rare but potentially fatal disorder both for mother and baby affecting 1:7000 to 1:16000 deliveries. [1]

Case History :

A 25 year old Hindu female primigravida with no comorbid conditions and previous normal antenatal check up presented at 36 weeks of pregnancy with yellowish discoloration of urine and sclera for 10 days, abdominal distension and pedal oedema for 5 days, and altered sensorium and decreased urine output for one day.

On admission patient was in altered sensorium, irritable, with sinus tachycardia- pulse rate of 110/min, Blood Pressure - 110/70 mmHg and Random Blood Sugar (RBS) - 56mg/dL. On examination sclera was icteric, there was bilateral pitting edema upto knees and moderate ascites. Respiratory and cardiovascular examination was normal. Laboratory investigations revealed altered liver and renal function tests with septicemia. Arterial Blood gas analysis showed metabolic acidosis. Laboratory investigations on admission were as follows:

Table 1 : Laboratory Investigations on admission

Hemoglobin	9.79 g/dL	ALP	227 IU/L
WBC count	23900/dL	S. Total Protein	5.8 g/dL
Platelets	1.63 lac/dL	S. Albumin	1.84 g/dL
PT INR	3.42	S. Creatinine	7.4 mg/dL
APTT	56 sec	S. Urea	154 mg/dL
SGPT	77 IU/L	S. Sodium	130 mEq/L
S. Total Bilirubin	18.6 mg/dL	S. Potassium	4.3 mEq/L
Direct Bilirubin	15.8mg/dL	Urine Routine/ Micro	3-4 pus cells , protein nil
S. LDH	125 IU/L	S. Uric acid	8.6 mg/dL

Patient was stabilized. 4 pints of Fresh Frozen Plasma (FFP) were transfused. Fetal cardiac activity was not recorded on fetal monitor. Ultrasonography (USG) confirmed intrauterine death. The spalding sign was absent suggestive of recent intrauterine death. Urgent obstretician consultation was done. Labour was induced and patient delivered still born male child.

Patient remained in altered sensorium post partum. One cycle of hemodialysis was done after delivery as patient had <100ml urine output in 24 hours. Patient had recurrent episodes of hypoglycemia. She was started on continuous dextrose drip, with higher antibiotics, diuretics and supportive treatment.

Differential diagnosis of Severe Pre-eclampsia, HELLP (Hemolysis, Elevated liver Enzymes, Low Platelet count), AFLP, Acute viral hepatitis, Cholestasis, Decompensated liver cirrhosis were considered. Viral markers [HBsAg (hepatitis B antigen), Antibodies against HCV (Hepatitis C Virus), HAV(Hepatitis A Virus), HEV(Hepatitis E Virus)] were negative. On urine routine microscopy there was no albuminuria, S.LDH was normal, platelet counts were normal. Ascitic fluid examination showed transudative fluid. Blood culture, urine culture, ascitic fluid culture revealed no growth. USG abdomen showed changes of fatty liver with moderate ascites thus confirming the diagnosis of AFLP.

Patient's sensorium started improving on the third day postpartum, urine output increased, ascites and pedal edema started resolving. On 10th postpartum day Renal Function test normalized alongwith declining bilirubin levels. Patient was discharged with S.bilirubin 4.3mg/dL which normalized after 3 weeks at follow up.

Discussion:

The differential diagnosis of jaundice during pregnancy includes cholestasis, cholelithiasis, viral hepatitis, pre-eclampsia with or without HELLP syndrome and AFLP.[2,3] Acute liver failure during pregnancy may be caused by fulminant viral hepatitis, drug-induced hepatic toxicity or AFLP. AFLP is more common in nulliparous with a male fetus and in 15% of cases there is a multifetal gestation. It usually occurs in late third trimester, with rare cases reported at 23-26 weeks. It is characterized by microvesicu-

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lar steatosis in the liver. The precise etiology of AFLP is not known but it is thought to be due to a mitochondrial dysfunction in the oxidation of fatty acids leading to an accumulation of fatty acids in hepatocytes. The infiltration of fatty acids causes acute liver insufficiency. Women who develop AFLP are more likely to have a heterozygous long-chain 3-hydroxyacyl-coenzyme A dehydrogenase (LCHAD) deficiency, found on the mitochondrial membrane and involved in the beta oxidation of long-chain fatty acids. This mutation is autosomal recessive. AFLP occurs if the fetus is homozygous for this mutation and thus is unable to metabolize fatty acids. [4,5]

Usually patient of AFLP presents with non specific complaints of malaise, nausea, vomiting, right hypochondriac pain, jaundice. In severe cases, the patient can present with multisystem involvement including acute renal failure, encephalopathy, gastrointestinal bleeding, pancreatitis and coagulopathy, hypoglycemia. Polydipsia and polyuria with transient diabetes insipidus have been reported. Signs of pre-eclampsia are found in 50% cases. Patients generally have leucocytosis (greater than 15 × 109/L), but a normal hematocrit. The prothrombin and partial thromboplastin times are both prolonged. Abnormalities in liver biochemistry include elevated serum aminotransferases (ie, AST/ ALT) levels of 300 U/L to 500 U/L, but the range has been reported to be from normal to 1000 U/L. Elevated serum aminotransferases may also be associated with raised serum ammonia, amino acid levels and lactic acidosis, elevated uric acid, hyperbilirubinemia (mainly conjugated without hemolysis) and hypoglycemia secondary to impaired hepatic glycogenolysis. Alkaline phosphatase may be elevated up to 10 times normal. The most striking feature of this syndrome is high level of bilirubin associated with moderate increase of transaminases. Blood urea nitrogen and creatinine may also be elevated, and acute renal failure may complicate severe cases.

Both ultrasound examination and computed tomography may demonstrate fatty infiltration of the liver. Liver biopsy is not necessary to establish the diagnosis. Liver biopsy reveals microvesicular steatosis with relative sparing of Zone 1(periportal).

Early diagnosis, prompt delivery and intensive supportive care are the cornerstones in the management of AFLP. Delivery is the definitive treatment of AFLP. Medical management of patients with AFLP is supportive. Orthotopic liver transplantation should be considered for those women with fulminant hepatic failure due to AFLP, who manifest signs of irreversible liver failure despite delivery and aggressive supportive care.

Maternal mortality rates from AFLP in the past were 75% but with prompt diagnosis and timely treatment mortality rate has decreased to approximately 18%. Deaths are usually secondary to sepsis, renal failure, circulatory collapse, pancreatitis or gastrointestinal bleeding. Neonatal mortality rates are usually higher around 60 to 85%.[6] These children are at risk of hypoglycemia, fatty liver, dilated cardiomyopathy, progressive neuromyopathy, and sudden infant death syndrome. Recurrence of AFLP in subsequent pregnancies can occur.

Conclusion:

AFLP is an uncommon life threatening complication of third trimester of pregnancy with variable presentation. It may occur rapidly and progression is unpredictable. High degree of suspicion is necessary for its diagnosis. It is recommended that patients with nausea, vomiting, epigastric pain and persistent jaundice in the third trimester with altered liver function tests, should be suspected for diagnosis of AFLP. Good maternal outcome is dependent on early diagnosis and immediate delivery.

References

1. Ahmed KT, Almashhrawi AA, Hammoud GM et al. Liver diseases in pregnancy: diseases unique to pregnancy. World J Gastroenterol 2013; 19(43);7639-46.
2. Sherlock's Diseases of the liver and biliary system- 12th edition, 30:602-614. Hay JE. Liver Disease in Pregnancy. Hepatology 2008; 47:1067-1076
3. L.L. Bellig, Maternal acute fatty liver of pregnancy and associated risk of LCHAD deficiency in infants, Advances in Neonatal Care, Vol 4(1):26-32, 2004.
4. Ibdah JA. Acute fatty liver of pregnancy an update on pathogenesis and clinical implications. World J gastroenterol 2006;12:7397.
5. Rajasri AG, Srestha R. Acute fatty liver of pregnancy- an overview. J Obstet Gynaecol Apr 2007; 27(3):237-240