Case Report

An Uncommon Case of Acute Fatty Liver of Pregnancy

Sanjay Prakash, Prashant Ramdas Wankhade, Krishna Priya Ganguly

Consultant Anaesthesiologist & Intensivist, Orchid Hospital, New Delhi, India.
Consultant Physician & Intensivist, Orchid Hospital, New Delhi, India.
Consultant Gynaecologist, Orchid Hospital, New Delhi, India.
Correspondence to: Sanjay Prakash, E-mail:drsanju_bell@yahoo.com

Received August 01, 2015. Accepted August 13, 2015

Abstract

Acute fatty liver of pregnancy is an uncommon life threatening condition presenting late in the third trimester of pregnancy. There is severe liver dysfunction, which needs to be differentiated from other conditions causing this, in pregnancy. Patients present with nonspecific complaints such as nausea, vomiting, malaise, fatigue, thirst, and altered mental state. If left undiagnosed and untreated it can progress to fulminant hepatic failure and death. Treatment is early delivery of baby and management of the patient on lines of liver failure in an intensive care unit.

KEY WORDS: Acute fatty liver of pregnancy (AFLP), haemolysis elevated liver enzymes and low platelets (HELLP) syndrome.

Introduction

Acute fatty liver of pregnancy (AFLP) is an uncommon potentially fatal disorder. Its incidence ranges from 1 per 7270 to 13,000 deliveries.[1] It carries a significant perinatal and maternal mortality.[2] It usually presents between 30th and 38th weeks of gestation, though reports of rare presentation in second trimester are also there.[3] It presents more frequently in primipara than multiparous women but can occur after multiple uneventful pregnancies and in subsequent pregnancies too.[4] Patients usually present with nonspecific complaints, such as headache, fatigue, nausea, and vomiting.[5] Diagnosis of AFLP is often delayed because of significant overlap in clinical and biochemical features with the haemolysis elevated liver enzymes and low platelets (HELLP) syndrome. Aetiology is not known clearly but some of the clinical and pathological features of AFLP are similar to those found in certain autosomal, recessively inherited disorders of fatty acid oxidation and hence it was suggested that AFLP may result from defects in β-oxidation of fatty acids.[6] We present here a case of AFLP in a second gravida patient that had a positive outcome because of early recognition followed by delivery of baby.

Access this article online	
Website: http://www.ijmsph.com	Quick Response Code:
DOI: 10.5455/ijmsph.2016.0108201583	

Case Report

A 33-year-old 31-week second gravida woman came with complaint of nausea, vomiting, and malaise since 10 days. There was no history of fever and headache. She was a known hypothyroid and was taking 100 μg of thyroxine daily. She had uneventful previous trimesters. On examination she was conscious, was icteric, and her heart rate was 80/min, blood pressure was 120/80 mmHg. Obstetric examination revealed less liquor and fetal bradycardia. Investigations on the day of admission showed raised liver function tests, rest all routine investigations were found to be normal. Her serum bilirubin was 7.08 mg/dL, conjugated bilirubin was 6.52 mg/dL. Aspartate aminotransferase (AST) was 1830 IU/L, alanine aminotransferase (ALT) was 2510 IU/L, and alkaline phosphatase was 1074 U/L. Viral markers were done. Hepatitis B surface antigen (HBsAg), HIV, VDRL, hepatitis C virus IgG (HCV), anti HEV (IgM) antibodies, anti HAV (IgM) antibodies were all negative. Coagulation profile was normal and serum thyroid stimulating hormone was 7.70. Ultrasound abdomen revealed oligoamnios and gall bladder wall thickening without gall stones. She was diagnosed provisionally as AFLP and patient was taken for emergency caesarean delivery in view of fetal distress. She was given spinal anaesthesia,

Table 1: Trend of liver function tests

Investigation	Day 1	Day 2 (Post Surgery)	Day 3	Day 4	Day 6	Day 8	Day 10
Serum bilirubin	7.08 mg/dL	6.23	5.73	2.37	8.57	8.32	5.4
Conjugated bilirubin	6.52 mg/dL	5.06	4.48	4.87	7.00	7.01	4.3
AST	1830 IU/L	2630	1290	370	180	180	120
ALT	2510 IU/L	1900	1260	930	340	270	160
INR	1.2	1.2	1.0	1.0		1.0	

AST, Aspartate aminotransferase; ALT, alanine aminotransferase, INR, International Normalization Ratio.

as her coagulation profile was normal and a healthy baby was delivered. Patient and baby remained stable after surgery and mother was kept in intensive care unit (ICU) for further observation and management on the lines of liver failure. Her serum bilirubin, ALT, and AST decreased gradually after delivery (Table 1).

Discussion

This pregnant patient came to hospital with jaundice in her third trimester. There are few conditions that cause jaundice in pregnancy, such as, cholestasis, cholelithiasis, viral hepatitis, pre-eclampsia with or without HELLP syndrome, and AFLP. In our patient, there was no cholestasis, cholelithiasis as evident through ultrasound abdomen. All viral markers were negative and patient had no manifestation of pre-eclampsia and HELLP syndrome throughout pregnancy. So a diagnosis of AFLP was made. Aetiology of AFLP is not clear. Deficiency of long chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) enzyme has been proposed as a cause of AFLP.[7] LCHAD is present in the mitochondrial membrane and is involved in the β-oxidation of long chain fatty acids. This gene mutation is recessive, if the fetus is homozygous for this mutation, it will be unable to oxidize fatty acids. These acids are passed to the mother who because of diminished enzyme function cannot metabolize the additional fatty acids leading to the accumulation of microvesicular fat in the hepatocytes. Typical symptoms are 1–2 weeks history of nausea, vomiting, abdominal pain, and fatigue. Progression from moderate to severe hypoglycemia, coagulopathy, marked decrease of antithrombin III activity, encephalopathy, and frank liver failure can rapidly ensue. Approximately 50% patients will also have signs of preeclampsia. DIC is seen in about 80–100% patients with AFLP as compared to 21% patients with HELLP syndrome.[8]

This condition is a medical, maternal, and fetal emergency that needs to be diagnosed and treated early, as complications could be fatal. Our case was unique as patient had not yet developed fatal complications mentioned above and was managed early. A confirm diagnosis can be made by liver biopsy. As the baby needs to be delivered out in an emergency, liver biopsy cannot be done routinely. In our case also, we could not perform a liver biopsy. Diagnosis was made by excluding other probable conditions. Ultrasound and computed tomography have been used but the sensitivity and specificity of these imaging studies are insufficient to make a definitive diagnosis and false negative results are common. Treatment is mainly delivery of baby and management of liver failure in ICU.

Conclusion

A patient with jaundice in pregnancy should always raise a red flag in the minds of physician/obstetrician. She should be investigated thoroughly keeping AFLP in mind. It is a medical, maternal, and fetal emergency. Early diagnosis and delivery of baby is the only definitive treatment along with the supportive therapy for liver failure. Post-delivery intensive treatment is always required for correction of complications such as coagulopathy and encephalopathy.

References

1. Ahmed KT, Almashhrawi AA, Rahman RN, Hammoud GM, Ibdah JA. Liver diseases in pregnancy: diseases unique to pregnancy. World J Gastroenterol 2013;19:7639–46.
2. Goel A, Jamwal KD, Ramachandran A, Balasubramanian KA, Eapen CE. Pregnancy-related liver disorders. J Clin Exp Hepatol 2014;4:151–62.
3. Monga M, Katz AR. Acute fatty liver in the second trimester of pregnancy. Prim Care Update Ob Gyns 1998;5:191.
4. Wilcken B, Leung KC, Hammond J, Kamath R, Leonard JV. Pregnancy and fetal long-chain 3-hydroxyacyl coenzyme A dehydrogenase deficiency. Lancet 1993;341:407–8.
5. Kamimura K, Abe H, Kawai H, Kamimura H, Kobayashi Y, Nomoto M, et al. Advances in understanding and treating liver diseases during pregnancy: A review. World J Gastroenterol 2015;21:5183–90.
6. Treem WR. Mitochondrial fatty acid oxidation and acute fatty liver of pregnancy. Semin Gastrointest Dis 2002;13:55–66.
7. Mackillop L, Williamson C. Liver disease in pregnancy. Postgrad Med J.2010;86:160–4.
8. Ibdah JA. Acute fatty liver of pregnancy: an update on pathogenesis and clinical implications. World J Gastroenterol 2006; 12:7397–404.

How to cite this article: Prakash S, Wankhade PR, Ganguly KP. An Uncommon Case of Acute Fatty Liver of Pregnancy. Int J Med Sci Public Health 2016;5:584-585

Source of Support: Nil, **Conflict of Interest:** None declared.