Acute fatty liver of pregnancy complicating a twin pregnancy

INTRODUCTION

Liver disorders complicate 3% of all pregnancies.[1] The spectrum of diseases range from mild anomalies to gross derangements. Failure in making correct diagnosis may result in increased morbidity and mortality for both mothers and foetuses.[2]

CASE REPORT

A 37-year-old primigravida with twin gestation at 34-week was admitted with epigastric discomfort, abdominal pain, malaise and vomiting. Diagnosed

with hypertension in third trimester, she was prescribed alpha methyldopa. Heart rate was 102/min and blood pressure was 170/90 mm Hg. Icterus was noticed later on the day. Foetal heart sounds were heard. Ultra sonography of abdomen showed fatty liver and twin pregnancy [Figure 1]. Investigations: haemoglobin - 10.7 g%, total count - 10,700/mm^3, platelets - 2.6 lakhs, urea - 11 mg/dl, creatinine - 1.4 mg/dl, uric acid - 5.3 mg/dl, lactate dehydrogenase - 442 U/L, peripheral smear negative for haemolysis. Hepatitis B surface antigen, hepatitis C virus and human immunodeficiency virus negative, prothrombin time test 48 s control - 14 s, International Normalised Ratio - 4.0, total bilirubin - 7.8 mg/dl (0.3-1.0 mg/dL), direct bilirubin - 4.0 mg/dl (0z, serum glutamic oxaloacetic transaminase - 316 U/L (0-35 U/L), serum glutamic-pyruvic transaminase - 313 U/L (0-35 U/L), alkaline phosphatase 974 (30-300 IU/L), total protein - 5.3 U/L, albumin - 2.8 U/L, globulin - 2.5 U/L, plasma ammonia - 93 meq/d, urine albumin++, cardiotocography showed evidence of foetal distress. A presumptive diagnosis of pre-eclampsia and/or acute fatty liver of pregnancy (AFLP) was made. Emergency caesarean section was planned under general anesthesia. She was given vitamin K and fresh frozen plasma (FFP). Induction by rapid sequence using thiopentone, followed by suxamethonium, trachea intubated using size seven endotracheal tube. Anaesthesia maintained with oxygen: Nitrous oxide, sevoflurane, fentanyl and atracurium. Mannitol and albumin were transfused to prevent hepatorenal syndrome. Twin babies delivered were healthy. Extubation and recovery were uneventful. Cryoprecipitate and FFP were transfused in post-anaesthesia care unit. Broad spectrum antibiotics started. Magnesium sulphate was continued at 2 g/h. Serum magnesium levels monitored 6th hourly were normal. On the second post-operative day, liver function deteriorated further. On the 3rd post-operative day, she had two episodes of tonic clonic convulsions and respiratory distress. Chest X-ray was suggestive of acute respiratory distress syndrome. Mechanical ventilation initiated in synchronized intermittent mandatory ventilation mode with pressure support. Antibiotic was changed to tobramycin based on culture and sensitivity report. Computed tomography scan brain and electroencephalography were normal. Repeat ultra sound scan of abdomen showed ascites with hyperechoic features suggestive of haemorrhagic ascites as the patient already had coagulopathy. There were no gall stones or dilatation of biliary tract. Central venous pressure was maintained between 8 and 12 mm Hg. Weaned off on the 7th day but non-invasive ventilation was continued for three more days. Hepatic function gradually improved. Viral markers were negative on repeated tests. Coagulopathy was corrected with a total of 32 units of FFP, 4 units of platelets and 12 units of cryoprecipitate. Patient gradually improved with supports and was shifted out from post-operative intensive care unit on the 20th day.

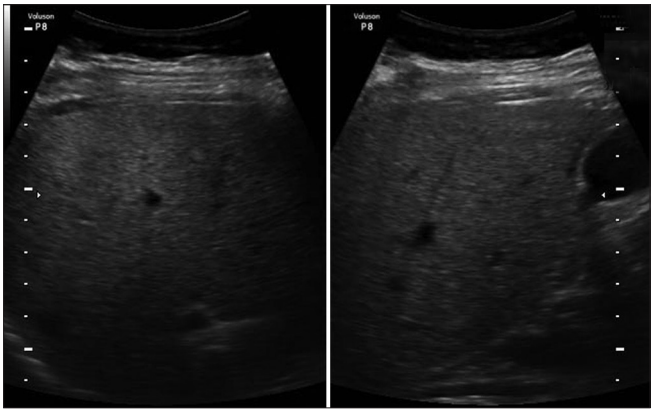

Figure 1: Ultrasonogram showing fatty liver

DISCUSSION

Pre-eclampsia, haemolysis, elevated liver enzymes, low platelets (HELLP) syndrome and AFLP are significant causes of maternal and perinatal morbidity and mortality. Several retrospective studies indicate an incidence of 1 in 13,000 pregnancies for AFLP compared to 1-6/1000 deliveries for the HELLP syndrome. AFLP is a life threatening condition with 18% maternal and 23% foetal mortality rate.

Pre-eclampsia occurs in 5% of all pregnancies usually in the second or third trimester.[3]

Symptoms of HELLP syndrome and AFLP appear in the third trimester.[4] In pre-eclampsia abnormal laboratory values include a 10-20 fold elevation in aminotransferases, elevations in alkaline phosphatase levels that exceed those normally observed in pregnancy, and bilirubin elevations of less than 5 mg/dL. Our patient had moderate elevation in the liver enzyme levels but serum bilirubin was markedly elevated, which was consistent with AFLP. Cause of AFLP is not known, many feel it to be a variant of pre-eclampsia. It has also been linked to inherited long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency in the baby. This is a rare autosomal recessive disorder. Both sexes can be carriers and when expressed, the baby is unable to metabolise some fatty acids and a

build-up can occur in the womb. Un-metabolised free fatty acids return from the baby, via placenta, to the mother's blood stream resulting in fatty liver.[5]

Criteria for diagnosis of AFLP include six or more of the following (Swansea criteria): Vomiting, abdominal pain, polydipsia/polyuria, encephalopathy, elevated bilirubin (>14 dmol/l), hypoglycemia (<4 mmol/l), elevated urate (>340 dmol/l), leukocytosis (>11 × 10^9/l), ascites or bright liver on ultrasound scan, elevated transaminases (aspartate aminotransferase or alanine aminotransferase >42 IU/l), elevated ammonia (>47 dmol/l), renal impairment (creatinine >150 dmol/l), coagulopathy (prothrombin time >14 s or activated partial thromboplastin time >34 s), microvesicular steatosis on liver biopsy.[6] Our patient had nine of the Swansea criteria, positive. The most striking feature of this syndrome is a high level of bilirubin associated with moderate increases of transaminases. Normal erythrocytic picture with normal platelets and no evidence of haemolysis also helped rule out HELLP.

Although a liver biopsy would have confirmed the diagnosis, it was not done due to coagulopathy. AFLP, characterized by microvesicular fatty infiltration of hepatocytes, is a disorder which is unique to human pregnancy.[7] It was described in 1940 and was initially thought to be universally fatal. The definitive management of AFLP is rapid delivery of the foetus and supportive care. Usually jaundice, liver dysfunction, and disseminated intra-vascular coagulation (DIC) may progress for 1-2 days after delivery but will then improve.[8] Before 1980, both the maternal and foetal mortality rates were about 85%,[9] and major causes were cerebral oedema, gastrointestinal haemorrhage, renal failure, coagulopathy, and sepsis. Although early caesarean section helped us to save both the babies we were unable to arrest the progression of the disease. Previously reported cases with AFLP had resulted in loss of foetus probably due to delayed delivery.[10]

Management of AFLP is mainly supportive. Fluid management, maintenance of glucose levels and correction of coagulopathy should be meticulously done. Foetus should be monitored with continuous cardiotocography. Early delivery of the foetus and maintaining maternal haemodynamic stability are the keystones.

The complications of AFLP require treatment after delivery, especially if pancreatitis occurs. Liver transplantation may be needed for mothers with severe DIC, those with rupture of the liver, or with severe encephalopathy.

CONCLUSION

The cornerstone of treatment in AFLP is early resuscitation of mother with meticulous monitoring, fluid management, coagulopathy correction, nutrition and mechanical ventilation, if needed. Early delivery of foetus improves the maternal outcome.

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