

Acute Fatty Liver in Pregnancy with Encephalopathy, Acute Kidney Injury, Disseminated Intravascular Coagulopathy and Acute Limb Ischaemia - A Case Report

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Abstract

Acute fatty liver in pregnancy (AFLP) is a rare but life threatening obstetric emergency. It can progress up to liver failure if not recognized and treated early. Generally pregnant mothers in third trimester and postpartum mothers are more prone to develop AFLP. 24 year old mother whose first baby was delivered by Emergency Lower segment Caesarean Section (Em LSCS) due to Lack of progression of labour presented in her second pregnancy at Period of Gestation (POG) of 37 weeks and 6 days with abdominal pain, nausea and vomiting and yellowish discoloration of sclera for 2 days duration. Unfortunately she developed acute limb ischaemia needing above knee amputation. Pathophysiology of Acute limb ischaemia may be an arterial embolus. Early detection and multidisciplinary management with Anaesthetic team, Intensive care team, Surgical team, Vascular surgical team, Obstetric team, Psychology team, physiotherapy team and social services section involved in helped the patient recover from this life threatening condition.

Keywords: Acute fatty liver; Pregnancy; Obstetric emergency

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Introduction

Acute fatty liver in pregnancy (AFLP) is a rare but life threatening obstetric emergency [1-3]. It can progress up to liver failure if not recognized and treated early. Generally pregnant mothers in third trimester and postpartum mothers are more prone to develop AFLP [1]. According to Royal College of Obstetricians and Gynecologists e-learning (2020), Incidence of AFLP is 1 in 7000-15000 pregnancies. Prim gravid women, elderly pregnant women, pregnant women with male fetuses, multiple pregnancies and pregnant mothers with low body mass index (BMI) are at a higher risk of developing AFLP [1-3]. AFLP is a clinical diagnosis with supporting evidence from laboratory investigations [3,4].

Pathophysiology of Acute Fatty Liver in Pregnancy is thought to be related to deranged metabolism of free fatty acids leading to micro vesicular fatty infiltration of the liver which can progress to hepatic failure [3]. Autosomal recessive genetic component appears to play a role [2]. Early delivery of the fetus is the definitive treatment of AFLP [1,2,4]. If not recognized and managed early, patients can develop complications like acute kidney injury,

coagulopathy and Severe Haemorrhage, fulminant hepatic failure and encephalopathy [3-5].

Case Report

A 24-year-old mother whose first baby was delivered by Emergency Lower segment Caesarean Section (Em LSCS) due to Lack of progression of labour presented in her second pregnancy at Period of Gestation (POG) of 37 weeks and 6 days with abdominal pain, nausea and vomiting and yellowish discoloration of sclera for 2 days duration. Fetal movements were satisfactory. Her pregnancy was a planned pregnancy with pre-conceptual Folic acid and Rubella vaccination. Her booking visit investigations were normal and the pregnancy was uncomplicated until this presentation at POG 37 weeks and 6 days. Her past medical history was unremarkable and the only past surgical experience was the Emergency Lower segment Caesarean Section done in previous pregnancy.

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On Examination she was icteric but not pale and afebrile. Blood pressure 120/60. Abdomen was soft and no hepatosplenomegaly detected. Fetal heart sounds were heard normally. Her investigations revealed Haemoglobin 12.2 g/dL, platelet count 76000, WBC 14.3×10^3 with neutrophils 67% and Lymphocytes 21%. Total bilirubin 15.8 mg/dL with direct fraction 8.24 mg/dL and indirect fraction 7.64 mg/dL. AST 119 U/L, ALT 151 U/L, ALP 313 U/L. CRP 6.9 mg/L, serum Creatinine 1.56 mg/dL, BU 58 mg/dL, ECG sinus tachycardia, CBS 58. Her CTG showed Variable decelerations. Ultrasound scan of abdomen showed acute parenchymal changes of liver. Diagnosis was made as acute fatty liver in pregnancy with fetal distress. EM LSCS done to deliver the baby urgently after optimizing the patient as the Cervix was not favorable. Liquor was Meconium stained and the did not cry at birth. Heavy uterine bleeding was noted and Intravenous Syntocinon 40 Units infusion, Intravenous Ergometrin 0.5 mg, Per Rectal misoprostol 800 ug and, Intravenous Tranexemic acid 1 g given. Intravenous Ceftriaxone 2 g stat, Intravenous Metronidazole 500 mg stat given prophylactically. Once the patient was stabilized haemodynamically she was transferred to a tertiary care hospital.

The patient was thereafter managed at Intensive Care Unit of the tertiary care Centre. On Post-operative day two she has been detected with intra-abdominal haemorrhage with altered clotting profile. Prothrombin time has been 36.1 sec (11-13.5) and INR 3.08 (0.8-1.1). Patient has been optimized and preceded with reopening and subtotal Hysterectomy. Patient has become confused and drowsy with examination findings in favour of encephalopathy. Hence the diagnosis has been made as Acute fatty Liver in Pregnancy complicated with hepatic encephalopathy, acute kidney injury and disseminated intravascular coagulation (DIC) using 'Swansea criteria'. The GCS has rapidly dropped needing incubation & ventilation. IV L-ornithine L-aspartate (LOLA) & Ursodeoxycholic acid have been given. 49 units of Red Cell Concentrates and 17 units of FFP, 16 units of platelets and 15 units of cryo precipitate have been transfused. 9 cycles of Total Plasma Exchange and 2 cycles of Continuous Renal Replacement Therapy (CRRT) have been completed. On post-operative day four she has developed acute limb ischaemia of Left Lower limb for which vascular surgical team of the tertiary care unit got involved.

Despite utmost efforts to preserve the left lower limb with vascular surgical procedures and fasciotomy, acute limb ischaemia has become worse. Therefore the patient had to undergo above knee amputation as a life-saving procedure. Continuous physiotherapy and gradual mobilization is done. Psychological and social

support has been provided to assist her cope up with challenges of life with above knee amputation of left lower limb. The patient was transferred back to our hospital on post-operative day 31. Repeat Ultrasound Scan after 8 weeks revealed improved Liver parenchyma. Other investigations revealed improving liver, renal functions and clotting profile.

Discussion

Acute Fatty Liver in Pregnancy (AFLP) though rare, is one differential diagnosis for jaundice during pregnancy. Other differential diagnoses for jaundice in pregnancy include acute viral hepatitis, gallstone disease intrahepatic cholestasis in pregnancy, autoimmune hepatitis, Pre-eclamptic live disease and HELLP Syndrome [1]. Our team could rule out acute viral hepatitis, gallstone disease, and intrahepatic cholestasis in pregnancy, autoimmune hepatitis rapidly using the history, examination findings and investigations. As pre-eclamptic live disease, HELLP Syndrome and AFLP share some of the features [4], it was a challenge to arrive at the definitive diagnosis. The prominent features that lead us to diagnose this patient as AFLP are jaundice, encephalopathy, coagulopathy, hypoglycemia, moderate rise of AST & ALT. Though live biopsy is the diagnostic investigations of AFLP it was contraindicated here due to coagulopathy. But generally the diagnosis of AFLP is clinical based on history, examination, laboratory investigations and imaging using "Swansea Criteria" [3].

We followed the definitive management of termination of pregnancy by urgent delivery of fetus [1-3], which was term with POG 37 weeks and 6 days. Except for some fetal distress which improved following initial resuscitation the new-born was devoid of significant complications. Typically maternal clinical condition is supposed to improve after delivery [1], but in this case maternal condition was persistently deteriorating with severe postpartum Haemorrhage and hepatic encephalopathy. Unfortunately she developed acute limb ischaemia needing above knee amputation [6-10].

Conclusion

Pathophysiology of acute limb ischaemia may be an arterial embolus. Early detection and multidisciplinary management with Anaesthetic team, Intensive care team, Surgical team, Vascular surgical team, Obstetric team, Psychology team, physiotherapy team and social services section involved in helped the patient recover from this life threatening condition.

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