Case Report

Acute fatty liver of pregnancy - a night mare

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Abstract

Introduction: Acute Fatty liver of Pregnancy (AFLP) is a rare and potentially life threatening obstetric emergency. The incidence of Acute Fatty Liver of Pregnancy is 1 in 7000 to 1 in 16,000. We report a case of AFLP in pregnancy in view of rarity and stormy progression and complications encountered during the management of the patient.
Keywords: Acute Fatty liver of pregnancy, Jaundice, disseminated intravascular coagulation, elevated liver enzymes.

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INTRODUCTION

Acute Fatty Liver of Pregnancy (AFLP) is a rare and life threatening complication, usually manifest in the late third trimester of pregnancy or early post partum period. The incidence is 1 in 7000 to 1 in 16,000 pregnancies.[1] It is common in primi gravida, pregnancies carrying male fetus and twin pregnancy. Maternal mortality is estimated to be 12.5-18% with a neonatal mortality rate of 7-66%.[2] The exact etiology is unknown but literature points to hepatic damage caused by defects in the activity of long chain 3 - hydroxyacyl coenzyme A dehydrogenase.[3] We are reporting a case of AFLP which had a stormy progression with multi systemic complications managed by a "multi disciplinary team approach" and had a good maternal and perinatal outcome.

CASE REPORT

A 22 years old primigravida, 39 weeks of gestation, conceived after ovulation induction presented with decreased appetite for 2 days, yellowish discolouration of eyes and swelling of legs with decreased urine output for 1 day and bleeding from the gums for 4 hours. There is no history of fever or itching. She is a known case of gestational diabetes, well under control with Tab. Metformin 500mg tid. On examination, there was no pallor, icteric with bilateral pitting pedal edema. Her BP recording was 130/80mmof Hg. Per abdomen uterus corresponded to term, relaxed, cephalic 3/5 palpable, fetal heart was good. Bishop score was 3, Initial laboratory parameters showed Hb of 11gm%, INR of 2.2, serum bilirubin of 10.58mg/dl with marginal elevation of liver enzymes, CBG of 50gm/dl, and platelet count of 3.55 lakhs/ dl and elevated serum Ammonia levels of 144mg/dl. Her renal function tests were normal. She was transfused 4 units of FFP. In view of unfavourable cervix, she was taken up for emergency LSCS. She delivered a live male baby of 3.08kg with APGAR of 8/10 and 9/10. There was yellowish discolouration of all the abdominal viscera. Bleeding was controlled with 20 units of oxytocin, injection carboprost 250µg IM and 1000mg of misoprostol rectally. Prophylactic Uterine Artery Embolisation (UAE) was done anticipating PPH in the same sitting and 28 F abdominal drains were placed. Intra operatively 6 units of FFP and 1 unit packed cells were transfused. Patient was shifted to intensive care unit in view of elective ventilation. The patient was extubated 20 hours later and her haemoglobin ranged from 3 gm% to 9gm% with replacement of blood products. On second post operative day, she developed decreased urine output and Acute Kidney Injury (AKI -II) was suspected. She was started on Continuous Renal Replacement Therapy (CRRT) for 24 hours and 10 sessions of hemodialysis later in the post operative period. In view of persistent collection in the drain and abdominal distension and drop in the haemotocrit, CT scan abdomen with angiogram was done; which revealed enlarged fatty liver, massive hemoperitoneum with bilateral pleural effusion. As the

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patient was desaturating, she was reintubated. Her laboratory parameters were monitored serially and extubated after 72 hours. The abdominal drain had to be removed as it was dislodged. By post operative day ten, bilirubin level reached a peak value of 31.3 mg/dl and the patient developed spiking temperature with diarrhoea. Blood culture sensitivity showed Klebsiella pneumonia and was treated with antibiotics as per the sensitivity. The serial ABG monitoring showed worsening of PO2 and chest X ray showed consolidation with pleural effusion. She was re intubated for the third time and continued with supportive care. As there was improvement in Oxygen Saturation, she was extubated after 72 hours. In view of persistent spiking temperature with discharge from the wound, CT scan abdomen was repeated which showed a normal size of liver, a large hematoma in the right lumbar region measuring 14x11x16 cm compressing the ureter with bilateral hydro ureteronephrosis (HUN) and moderate hemoperitoneum. However there was no active bleeder. A pig tail inserted under ultrasound guidance did not drain adequately. Hence with the suspicion of infected intra abdominal hematoma, not responding to this management, it was decided for laparotomy under general anesthesia on 33rd post operative day. Intra operatively, there were loculated collection of clots and hemoperitoneum of two litres, the same was drained and 28 F intra abdominal drain was placed. Patient's general condition and laboratory parameters showed improvement. The abdominal drain was removed on post operative day 10 and sutures on day 15. The patient was discharged on post operative day 45 of the first surgery with bilirubin level of 5.14mg/dl and Hb of 10 gm%. She was transfused with 110 units of blood and blood products which included packed cells, fresh frozen plasma, platelets, cryoprecipitate and fibrinogen. The patient was followed up after 2 months with gross improvement in clinical and laboratory parameters.

DISCUSSION

AFLP is a rare, advanced gestational complication which occurs around 32 -38 weeks. Since AFLP shows atypical presentation with abrupt evolution of complications resulting in multi organ dysfunction, it needs emergency referral and prompt treatment. The diagnosis of AFLP is made on the basis of clinical and laboratory criteria[4]

1. Patients with the symptoms of nausea, vomiting, epigastric pain, polydipsia / polyuria, jaundice and abnormal liver function in third trimester of pregnancy.
2. Characteristic laboratory examination of elevated serum bilirubin level, elevated uric acid, hypoglycaemia, leukocytosis, elevated transaminase.
3. Ultra sound imaging showing fatty liver.

Even our patient presented with decreased appetite, jaundice, elevated liver enzymes, serum Ammonia and uric acid, low glucose levels; with fatty liver on imaging and hepatitis screening being negative, a diagnosis of AFLP was considered. Delivery is the definitive treatment[4] of AFLP as the condition generally improves soon after the delivery. All patients should be hospitalised as soon as the diagnosis is made. If vaginal route is not favourable, caesarean section should be considered. As the risk of post partum hemorrhage is high, hysterectomy and UAE should be considered. Hence prophylactic UAE was done in our patient by which we could avoid hysterectomy in our patient. In the post partum period, they should be monitored for Coagulopathy and liver cell failure. The temporal profile of the disease leads to coagulopathy, fulminant hepatic failure, multi organ dysfunction, repeated hypoglycaemia, rapid progression to coma and ultimately death. Our patient even though she rapidly progressed to coagulopathy, renal insufficiency and hepatic inadequacy, timely early diagnosis, prompt intervention by multidisciplinary team approach with adequate supportive care in a tertiary care centre was the key to a positive maternal and perinatal outcome.

CONCLUSION

Though AFLP is an uncommon life threatening complication of third trimester of pregnancy, early diagnosis, prompt delivery, collaborative management in ICU and a multi disciplinary team approach are the key for a good maternal and perinatal outcome.

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