Case Report

Indian Journal of Obstetrics and Gynecology
Volume 8 Number 4, October–December 2020
DOI: http://dx.doi.org/10.21088/ijog.2321.1636.8420.13

Acute Fatty Liver of Pregnancy - A Fatal Complication in Pregnancy: Case Series

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How to cite this article:
Pooja Gupta, S K Singh. Acute Fatty Liver of Pregnancy - A Fatal Complication in Pregnancy: Case Series. Indian J Obstet Gynecol. 2020;8(4):275–278.

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Abstract

Introduction: Acute fatty liver of pregnancy (AFLP) is a rare, but potentially fatal condition, characterized by hepatic failure typically in the third trimester of pregnancy that is associated with multiorgan involvement resulting in several clinical and laboratory abnormalities.

Aim and Objectives: To study the course of acute fatty liver of pregnancy which will help later to early diagnose, prompt delivery, and providing supportive care to improved maternal morbidity and mortality rates.

Materials and Methods: The authors report an observational case series of patients who developed acute fatty liver of pregnancy and managed in critical care in a year that illustrated the compound challenges in recognition, monitoring, and management. One of the cases has been imprinted with maternal mortality because of deterioration of the condition leading to sepsis along with multiorgan dysfunction syndrome.

Results: A total of three cases of acute fatty liver of pregnancy were found in a year. Nausea, vomiting, and anorexia were the most common symptoms among all. Jaundice and ascites were the most common findings. Deterioration of the condition can lead to complications like sepsis, Disseminated Intravascular Coagulation, multiorgan failure, and death.

Conclusions: Early detection, delivery, and supportive care are sufficient to reduce fatality but in cases of deterioration of patients' condition despite proper management can lead to multiple complications and ultimately mortality. Regular antenatal health check-ups can help in early detection.

Keywords: Acute fatty liver of pregnancy; Hepatic failure; DIC; Multiorgan failure; Sepsis.

Introduction

Acute fatty liver of pregnancy (AFLP) is a rare but potentially fatal condition for both mother and baby, as often the diagnosis is delayed. It is characterized by the accumulation of micro vesicular fat that "crowds out" normal hepatocytic function. It has an incidence of 1 in 10,000–16,000 pregnancies.[1] It occurs in the latter part of pregnancy and may be part of a spectrum of disorders related to pre-eclampsia.

Most suggested etiopathogenesis of acute fatty liver of pregnancy is the deficiency of the enzyme, long-chain 3-hydroxy acyl-CoA dehydrogenase (LCHAD) results in accumulation of medium- and long-chain fatty acids. It is an autosomal recessive disorder and the heterozygous LCHAD deficiency has been identified in some women with AFLP.[2]

Clinical findings in AFLP vary because it may occur with varying degrees of clinical severity and in conjunction with other third trimester symptoms, making an early diagnosis of AFLP difficult. Patients often present with nonspecific symptoms such as anorexia, nausea, vomiting, malaise, fatigue, headache, and abdominal pain. On physical examination, the patient is usually febrile and jaundice, which is very common and eventually occurs in more than 70% of patients with AFLP as the condition progresses.[3] Tenderness in

the right upper quadrant or mid epigastric area may be present.[4] In severe cases, the patient can present with multisystem involvement including acute renal failure, encephalopathy, gastrointestinal bleeding, pancreatitis, and coagulopathy.

The diagnosis of AFLP depends mainly on clinical presentation and laboratory findings. Swansea criteria is one of which 6 or more findings confirm the diagnosis of AFLP:

Clinical	Vomiting, Abdominal pain, Polydipsia/ polyuria, Encephalopathy.
Biochemical	Bilirubin >14 µmol/l
	AST/ALT >42 IU/l
	Ammonia >47 µmol/l
	Renal Urate >340 µmol/l
	Creatinine >150 µmol/l
	Endocrine Glucose < 4 mmol/l
	Leucocytosis >11 x109 /l
	Coagulopathy-PT >14 secs or APTT >34 secs (often with Plt count >100 x1012/l)
Radiological	Abdominal USS bright Liver echo texture / ascites
Histological	Biopsy Micro vesicular steatosis

Early diagnosis, prompt delivery, and intensive supportive care are the Triad in the management of AFLP. Maternal stabilization should be achieved before delivery, which includes airway management, treatment of hypertension, and correction of hypoglycaemia, electrolyte, and coagulation abnormalities. Maintenance of Intravenous fluids and blood products, as well as the frequent assessment of maternal vital signs, and evaluating the changes in mental status are all crucial. After stabilizing the mother, the next step is to deliver the fetus. Vaginal birth is considered the best approach; however, caesarean birth is often performed because of rapidly deteriorating maternal-fetal conditions.

Case Discussion

Case 1

20 years old, Primigravida with 37 weeks pregnancy came with chief complaints of decreased fetal movements since 1 day, vomiting since 2–3 days, fever on and off since 4–5 days. Her vitals on admission: pulse – 68/min, BP- 100/60 mmHg, icterus present. On per abdomen examination -uterine fundal height 32 weeks, Fetal heart sounds were absent, Ascites present .Her obstetrics ultrasonography findings were suggestive of Intrauterine death of 31weeks and ultrasonography of abdomen and pelvis suggestive of heterogeneous echotexture of the liver. Her laboratory findings were random blood sugar – 69 mg/dl , Haemoglobin – 13.6 g/dl, TLC- 26,300/cumm , platelet- 1.75 L , Sr. Albumin- 2.7g/dl, Sr. Total Bilirubin – 13.2 mg/dl , Sr. Direct Bilirubin – 12.56 mg/dl , SGOT- 241 IU/L, SGPT- 363 IU/L , PT-90 sec, INR- 7.7 , APTT-118 , LDH- 1799 , Creatinine – 2.6 mg/dl, ammonia – 110. Viral markers (HbsAg, HAV, HEV, HCV)- non-reactive. The patient was managed in the critical care unit. She was delivered vaginally after four FFP transfusion. There was a marked increase in ascites noted on day 3 postpartum, ascitic tapping done thrice. Multiple hypoglycemic episodes were noted which were corrected with 25% dextrose and multiple episodes of fever for which blood culture showed staphylococcus haemolyticus growth. Broad-spectrum antibiotics started (Piperacillin, Tazobactam, Meropenem). Patient Mortality occurred due to multiorgan dysfunction syndrome (liver failure, AKI, DIC, Sepsis).

Case 2

28 years old, Primigravida with 37 weeks 6 days pregnancy came in latent labor with chief complaints-pain in abdomen for 4 hours. On examination, Pulse- 90/min, BP- 110/70 mmHg, icterus present, pedal edema present, Per abdomen examination showed Uterine fundal height – 36 weeks, mild contractions, Fetal heart sounds regular. On per vaginal examination - Cervix- 2cm dilated, 30% effaced , station – 3. Her laboratory investigations - Hb- 13g/dl, TLC- 16100/cumm , platelet- 2.10 L , BSL R – 65 mg/dl , Sr. Total Bilirubin- 10.11mg/dl, Sr. Direct Bilirubin – 9.54 mg/dl , SGOT-133 IU/L , SGPT- 249 IU/L , PT- 10.11 sec , INR – 1.69 , Ammonia- 104 , viral markers(HbsAg , HAV , HEV , HCV)-Non-Reactive and Ultrasonography of Abdomen and pelvis showed liver – coarse echotexture of liver. The patient was taken for emergency LSCS done due to fetal persistent bradycardia. She was managed in the critical care unit post-cesarean, 8 FFP transfusion done. She had multiple hypoglycemic episodes which were corrected with 25 % dextrose solution, broad-spectrum antibiotics (Piperacillin and Tazobactam) started. The decreasing trend of bilirubin and liver enzymes was noted in subsequent days. On the 5th Postoperative day, she was shifted to Obstetric HDU. She recovered and was discharged on the 11th post-operative day.

Case 3

29 years old, G2P1L1 with 38 weeks 1 day of pregnancy came with Premature rupture of membranes for 3 hours. On general examination, Pulse -98/min, BP-120/80 mmHg, icterus present. on per abdomen examination, uterine height was 36 weeks, cephalic presentation, persistent fetal heart rate drops up to 80–90 beats /min noted. On per speculum examination, thick meconium-stained liquor was noted. Her laboratory findings were random blood glucose - 55 mg/dl , Hb- 12.1 g/dl, TLC- 21300/cumm, platelet-1.45 L, Sr. Total Bilirubin- 14.39 mg/dl, Sr. Direct Bilirubin -12.57 mg/dl, SGOT- 104 IU/L, SGPT-199 IU/L, PT- 26.7 sec, INR-2.23, Sr. Creatinine-1.06, LDH-938, Ammonia -90. The patient was taken for an emergency cesarean section and was kept in the critical care unit later. Post-cesarean ultrasonography showed a coarse echotexture of the liver. She developed multiple hypoglycemic episodes which were corrected with a 25 % dextrose solution. Four FFP transfusion done post-cesarean. Broad-spectrum antibiotics were given to her (meropenem, tigecycline). Serial monitoring of her liver function tests and coagulation profile showed a decreasing trend. On the 6th postoperative day, she was shifted to Obstetric HDU. The patient improved clinically and laboratory parameters came down to normal values. The patient was discharged on the 15th post-operative day.

Discussion

In our hospital, Three cases of Acute Fatty Liver Of Pregnancy were treated during September 2019-September 2020. Two patients were primigravida of which one undergoes vaginal delivery while the other underwent cesarean section. The remaining one patient was multigravida who underwent a cesarean section. In cases that have been discussed above, two patients recovered completely while one patient lends up into maternal mortality. All three patients were diagnosed with AFLP on the basis of fulfilling the Swansea criteria and there was no need for doing liver biopsy which is considered as the gold standard test for diagnosis of AFLP.

Acute Fatty liver of pregnancy (AFLP), is a disease entity unique to pregnancy, which is usually encountered towards the third trimester.[5] The clinical evidence is sufficient to diagnose AFLP in many cases - Swansea criteria. Diagnosis can be confirmed by a liver biopsy but it is not done routinely. Ultrasound scanning remains the imaging modality of choice as it is both safe and as well as convenient.

As a result of early diagnosis and intervention, the worldwide maternal mortality recorded as 100% in the past, have now reached rates <10%.[6] The disease, if not intervened early, maybe complicated with upper gastrointestinal bleeding due to altered coagulation profile, AKI, infection, pancreatitis, or hypoglycemia.[7]

Early diagnosis, prompt delivery, and intensive supportive care are the cornerstones in the management. Maternal stabilization should be achieved before delivery, which includes airway management, treatment of hypertension, and correction of hypoglycemia, electrolyte, and coagulation abnormalities. The delivery of the fetus is the next step. Vaginal birth is probably the best approach if tolerated. Cesarean birth is often performed because of rapidly deteriorating maternal-fetal status.[8]

Recurrence risk in subsequent pregnancies is 25% with a mother carrying a homozygous mutant or compound heterozygous fetuses, it is uncommon and only a few cases have been documented. However, this may be an under-representation, because many women may refrain from having further pregnancies after the first occurrence. Therefore, affected women should still be informed, counselled , and tested, along with their infants who may be affected, for LCHAD deficiency.[9,10] If the patient decides to be pregnant again, she should be closely monitored for any early signs of acute fatty liver.

Conclusion

Through this case series, I would like to reemphasize the grave nature of this disease entity which is unique with pregnancy. Early detection of the condition, early delivery, and prompt supportive care is sufficient to reduce morbidity and mortality but in cases of deterioration of the patient's condition despite proper management can lead to multiple complications and ultimately mortality.

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