

Acute Fatty Liver of Pregnancy - A Rare but Fatal Obstetric Complication

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Abstract: *Acute Fatty Liver of Pregnancy (AFLP) is a rare, catastrophic disease affecting women in the third trimester of pregnancy or in the post - partum period. It is usually a diagnosis of exclusion and requires a strong index of suspicion for a timely diagnosis and prompt intervention. Case Summary: A 39 yr old primigravida, IVF conceived at 29 wks period of gestation was referred from Indira IVF center ivojaundice. She had yellowish discoloration of eyes and urine since 3 days. Her AST/ALT was 2911/1889. There was no history of fever, vomiting and diarrhoea. Due to deteriorating maternal condition, her EmLSCS was done. Intra - op her liver was shrunken. Liver biopsy was taken which was suggestive of acute fatty liver. In post op period, pt was in ICU for 8 days, but she recovered. Conclusion: There should be a high index of suspicion of this condition in women presenting with jaundice in pregnancy. Delay in diagnosis is associated with increased maternal morbidity and mortality.*

Keywords: Acute fatty liver of pregnancy, Jaundice

1. Background

AFLP is a disease of the third trimester that is unique to human pregnancy and was described by Sheehan in 1940. The condition was associated with high mortality rates but this has improved because of early diagnosis and prompt delivery of the fetus. The approximate incidence of AFLP is 1: 7, 000 to 1: 20, 000. Conditions unique to pregnancy that cause liver dysfunction include intrahepatic cholestasis of pregnancy (ICP), pre - eclampsia, HELLP (hemolysis, elevated liver enzymes, low platelet count) syndrome and AFLP. While ICP and pre - eclampsia are frequently seen, AFLP is rare and potentially life - threatening. The pathogenesis of AFLP remains unclear but there is emerging evidence of the genetic basis of AFLP where defective mitochondrial fatty acid beta - oxidation in the fetus is implicated in some cases of AFLP. We present a case where early diagnosis and prompt management finally saved the pts life.

2. Case Report

A 39 yr old primigravida, IVF conceived with twin pregnancy presented to our hospital after being referred from Indira IVF centre. She had a history of yellowish discoloration of eyes and urine for 3 days, no history of fever, diarrhoea and vomiting. She had no history of travel to malaria endemic area. She had no prior history of itching. She was diagnosed with gestational diabetes 1 month back,

and was on diabetic diet and tab metformin. HIV status was negative. She had no chronic illnesses and no history of PCM, aspirin, sodium valproate intake. Her BP and pulse at the time of admission 106/68mm of Hg and 92/min respectively. She was icteric. Respiratory and cardiovascular examination were normal. The abdomen was soft and there was no hepatomegaly or splenomegaly. Fundal height was around 32 wks and both FHR were normal. She was concious and well oriented to time, place and person. Patient was admitted and all investigations were sent. Her Hb - 13.1, TLC - 15, 300, PLT - 3.17 lakhs, total Bil - 16.3, OT/PT - 1244/1946, ALP - 447, PT - INR - 2.81, Na+/K+ - 138/4.3. All her viral markers were negative. On day 2 of admission, her clinical condition deteriorated and she developed tachypnoea. Her Em Lscs was done under GA due to suspicion of acute fatty liver of pregnancy. Twin preterm female babies were delivered and were sent to nursery. Intra - op Ecchymosis were present on anterior parietal peritoneum. Liver was shrunken. With the help of surgeons, liver biopsy was taken. 4 unit FFPs were transfused intra operatively. She was not extubated in post op and was shifted to ICU for better care. On post op day 2, her total bilirubin increased to 19.3, and OT/PT was reduced (894/1023), Na+/k+ - 134/6.0. She also poor respiratory efforts and was unconscious, Her serum ammonia was 196.7. Triple I. V antibiotics were given. Her bilirubin was not reducing. so even liver transplant was considered. Her liver biopsy report was collected which was suggestive of fatty changes.

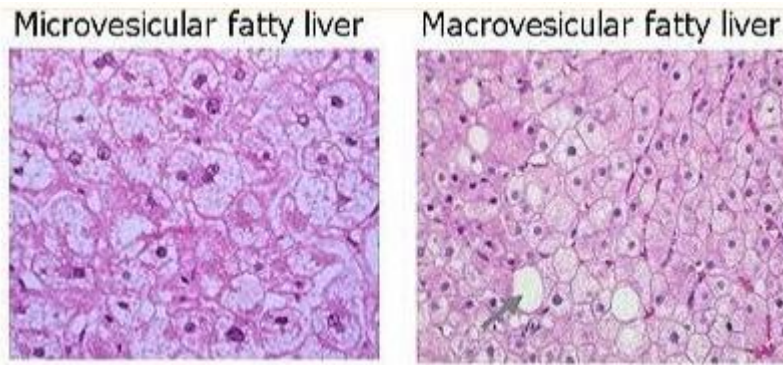


Figure 1: Fatty changes in liver

Gradually due to supportive intensive care, her bilirubin reduced to 11, OT/PT - 600/700, and coagulation profile came to normal. She was finally extubated and her general condition also improved.

3. Discussion

This case highlights the importance of a high index of suspicion of the condition (AFLP) in women presenting with jaundice in pregnancy. Other differential diagnoses of jaundice occurring during pregnancy include viral hepatitis, preeclampsia, cholelithiasis and intrahepatic cholestasis of pregnancy. The clinical presentation and laboratory findings are vague and non-specific and pose a diagnostic challenge. It is important to always consider life threatening differentials which may require prompt delivery and intensive care. While HELLP syndrome and AFLP usually complicate third trimester of pregnancy, HELLP syndrome (1 in 5000) is seen more frequently than AFLP (1 in 13000). In our case BP was always normal, there was no evidence of hemolysis and platelets were also normal, Urinalysis was negative for proteinuria, viral markers were also negative. Liver biopsy confirmed the diagnosis. Viral hepatitis also presents with jaundice but is characterized by a generally unwell patient with fever, nausea, vomiting with markedly elevated aminotransferases. Patients with intrahepatic cholestasis of pregnancy commonly complain of pruritus and their serum bilirubin levels do not usually exceed 6mg/dl. Ingestion of drugs and herbal remedies that could lead to hypoglycemia were ruled out from history. Patients with cholelithiasis, in addition to jaundice, also have pain in the right upper quadrant as well as fever and an ultrasound scan aids in the diagnosis. Cholelithiasis and viral hepatitis may occur at any time during pregnancy unlike AFLP which is usually diagnosed in the third trimester as noted earlier. Sepsis was unlikely as the patient had no tachycardia or hypotension and remained normothermic. Other differential diagnoses were excluded in our case based on the symptoms, timing of the presentation and investigations that were available.

Our patient had a marked rise in bilirubin with a fall in liver enzymes in the post-op period making us highly suspicious of liver failure, but with supportive intensive management she gradually improved.

The definitive management of AFLP is rapid delivery of the fetus and supportive intensive care. As seen in our case that, jaundice, liver dysfunction and coagulopathy progressed for

6 days following delivery, but gradually improved. The histological features were suggestive of a diagnosis of AFLP. Our patient presented in early 3rd trimester and even after delivery, her condition worsened for initial few days.

4. Conclusion

AFLP is a rare, life-threatening complication of third trimester which requires a high index of suspicion for early diagnosis. Urgent delivery and maximum supportive care should be instituted to prevent poor outcomes.

Abbreviations:

AFLP: Acute fatty liver of pregnancy;
ALP: Alkalinephosphatase;
ALT: Alanineaminotransferase;
ANC: Antenatalcare;
AST: Aspartateaminotransferase;
HELLP: Haemolysis, elevated liver enzymes, lowplatelets;
HIV: Human immunodeficiency virus;
ICP: Intrahepatic cholestasis of pregnancy;
PLT: platelets;

Conflicts of Interest:

The authors declare that they have no conflicts of interest.

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References

- [1] Sheehan H. The Pathology of Acute Yellow Atrophy and delayed chloroform poisoning. An International Journal of Obstetrics and Gynecology.1940; 47 (1): 46–62.
- [2] Vora KS, Shah VR, Parikh GP. Acute fatty liver of pregnancy: a case report of an uncommon disease. Indian J Crit Care Med.2009; 13 (1): 34–6.
- [3] Ibdah JA. Acute fatty liver of pregnancy: an update on pathogenesis and clinical implications. World J Gastroenterol.2006; 12 (46): 7397–404.
- [4] Pereira SP, O'Donohue J, Wendon J, Williams R. Maternal and perinatal outcome in severe pregnancy -

- related liver disease. *Hepatology* (Baltimore, Md.1997 Nov; 26 (5): 1258–1262.
- [5] English N, Rao J. Acute fatty liver of pregnancy with hypoglycaemia, diabetes insipidus and pancreatitis, preceded by intrahepatic cholestasis of pregnancy. *BMJ case reports*.2015; 15: 2015.
- [6] Bacq Y, Riely CA. Acute fatty liver of pregnancy: the hepatologist's view. *Gastroenterologist*.1993; 1 (4): 257–64.
- [7] Usta IM, Barton JR, Amon EA, Gonzalez A, Sibai BM. Acute fatty liver of pregnancy: an experience in the diagnosis and management of fourteen cases. *Am J Obstet Gynecol*.1994; 171 (5): 1342–7