International Journal of Gynaecology and Obstetrics Sciences
www.gynaecologyjournals.com
Online ISSN: 2664-9012, Print ISSN: 2664-9004
Received: 18-09-2021, Accepted: 02-10-2021, Published 19-10-2021
Volume 3, Issue 1, 2021, Page No. 18-20

Acute fatty liver of pregnancy and its outcome

Veena M Vernekar[1], Lakshmi A[1], Razıya[2], Ashadevi[3], Ashok Kumar K[4], Sreelatha S[4], Gurudath GS[5]

[1] Juniour Resident, Department of Obstetrics and Gynaecology, ESIMC PGIMSR, Rajajinagar, Bangalore, Karnataka, India

[2] Department of Obstetrics and Gynaecology, ESIMC PGIMSR, Rajajinagar, Bangalore, Karnataka, India

[3] Assistant Professor, Department of Obstetrics and Gynaecology, ESIMC PGIMSR, Rajajinagar, Bangalore, Karnataka, India

[4] Professor, Department of Obstetrics and Gynaecology, ESIMC PGIMSR, Rajajinagar, Bangalore, Karnataka, India

[5] Assistant Professor, Gastroenterologist, Department of Surgery, ESIMC PGIMSR, Bangalore, Karnataka, India

Abstract

Acute fatty liver of pregnancy is a rare, potentially fatal complication that occurs in the third trimester or early postpartum period.[1]

We are reporting a case of, 29 year old Primigravida with IVF conception with DCDA twin gestation who presented at 22 nd week of gestation with complains of abdominal pain and vomiting. Her Liver enzymes was raised. Ultrasound abdomen showed Acute Fatty changes in Liver and hepatomegaly, diagnosed as Acute Fatty Liver of pregnancy. Thereafter Gastroenterology opinion was taken. Patient was followed up antenatally, serial Liver function tests was done, her liver enzymes were normalised. Then at 33 week of gestation, she complained of leaking per vaginum. Emergency LSCS was done in view of DCDA Twin with Preterm Premature Rupture of Membrane. Both twin babies were shifted to NICU for Preterm management. The mother was stable postnatally. Liver function test was normal. Suture removal was done on post op day 7 and patient was discharged.

Keywords: acute fatty liver, pregnancy, DCDA twin, PPROM, emergency LSCS

Introduction

Acute Fatty Liver of Pregnancy (AFLP) though rare, is an obstetric emergency, which carries a high risk of maternal and perinatal mortality. It has an incidence of approximately one in 10,000 to 15,000 pregnancies [1]. AFLP was first described by Sheehan in 1940 as 'acute yellow atrophy of liver' [2]. But still the exact etiopathogenesis has not yet been clearly understood [1].

It usually occurs towards the latter part of pregnancy or in early post-partum period. But Monga and Katz reported a case of AFLP as early as in 22 weeks of gestation [3].

Another case reported by Suzuki *et al*, at 23 weeks of gestation [6]. Currently with prompt diagnosis and early management, the maternal and perinatal mortality have greatly decreased to approximately 18% and 23% respectively. The risk factors for developing AFLP are- primiparity, multiple gestation, male fetus sex, advanced maternal age, high body mass index, preeclampsia, prior episode of AFLP. [5]. Clinical presentation of AFLP is usually with non-specific and vague symptoms, hence a high index of suspicion is required especially if a pregnant women presents with symptoms of anorexia, nausea, unremitting vomiting, weight loss, malaise, head ache, abdominal pain, jaundice. Hepatic tenderness with hepatomegaly may or may not be present. The important differential diagnosis are fulminant viral hepatitis, severe HELLP syndrome, severe preeclampsia, cholestasis of pregnancy, gall stone disease, Budd Chiari syndrome etc. Prompt diagnosis and early management are the key measures for optimal maternal and fetal outcome in AFLP.

Case Report

A 29 year, primigravida conceived through *In Vitro* fertilisation with DCDA twin gestation presented at 22 weeks of gestation with nausea, vomiting, abdominal pain and malaise. She was normotensive with no other co morbidities. All routine investigations was done. She had elevated Liver transaminases levels [AST- 383 U/L; ALT- 388U/L]. Platelet count and coagulation profile was normal. Total /direct bilirubin level was normal. No proteinuria. Negative for Hepatitis serology. No chronic illness or drug abuse history.

The abdominal ultrasound showed a brilliant and bright river echo-texture with acute fatty changes in liver and hepatomegaly.

A diagnosis of Acute Fatty Liver of Pregnancy was made. Physician and Gastroenterologist opinion was taken. Serial Liver function tests were done, liver enzymes was eventually under normal limit. Patient was advised for regular antenatal checkups.

Then at 33 weeks of gestation, she complained of leaking per vaginum since one hour.

On examination vitals were stable , no icterus, no pedal edema , Per abdomen- uterus was irritable , both twins fetal heart sound was good, Per speculum examination revealed clear active leak, Per vaginal examination- Cervix was one cm dilated, minimally effaced, membranes absent, clear leak was present.

Emergency LSCS was done in view of DCDA twin with Preterm Premature Rupture of Membranes and extracted twin male babies of weight 2 kg and 1.6 kg. Both the twin were shifte to NICU for preterm management. The mother was stable postpartum. Hemoglobin level was 7g/dl, so one pint of packed red blood cell transfusion was done. Liver functions test were normal. Physician opinion was taken and advised Tab. Ursodeoxycholic acid 300mg BD and Syrup

Lactulose 15 ml HS.
Suture removal was done on post op day 7 and patient discharged.

Discussion

Acute Fatty Liver of Pregnancy, is a disease entity which is unique to pregnancy, though rare has significant morbidity and mortality. It generally occurs in third trimester or early postpartum. The pathogenesis is still poorly understood but defects in fatty acid metabolism during pregnancy appear to play role. Approximately 20% of AFLP is associated with fetal Long Chain 3-hydroxy acyl CoA Dehydrogenase deficiency (LCHAD) [5]. It is one of the enzyme involved in fatty acid oxidation, it catalyses the step in beta oxidation of mitochondrial fatty acid. In fetus homozygous for LCHAD deficiency, the feto-placental unit cannot perform this step, so the level of long chain fatty acid increases and enter maternal circulation which accumulates in maternal liver resulting in toxic effects. The most common mutation associated with AFLP is homozygous G1528C mutation [5].
There is progressive lipid accumulation within the hepatocytes with micro and macro vesicular steatosis of parenchymal cells, especially involving he periportal area. The liver grossly is soft, small yellow. If the disease process continues, it would eventually lead to atrophy of liver cells. Even kidney, Pancreas, Brain, Bone marrow may show micro vesicular fat infiltration. The usual laboratory investigation abnormality seen in AFLP include elevation in aminotransferase levels (AST and ALT) ranging from 5 to 10 times the upper limit of normal, elevated bilirubin, serum creatinine, serum uric acid and ammonia levels, Thrombocytopenia, low serum glucose, prolonged Prothrombin Time, low fibrinogen levels and leucocytosis.
Ultrasound Abdomen is the safest and convenient imaging modality but shows non-specific changes - brilliant bright echo texture of liver with fatty infiltration. MRI or CT abdomen can be done. Liver biopsy confirms the diagnosis but is rarely indicated due to accompanying coagulopathy with risk of intra-abdominal bleeding. Histopathology shows micro and macro vesicular steatosis of parenchymal cells with sinusoidal dilatation, with no interface hepatitis (Figure 1).

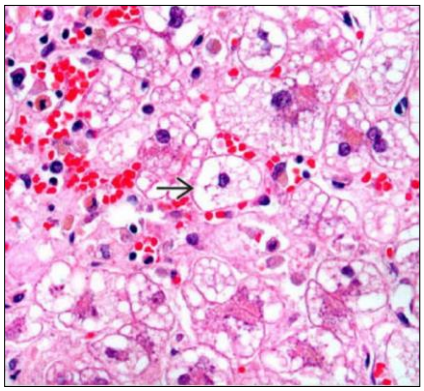

Fig 1: Micro vesicular steatosis of liver parenchymal cells. Arrow- The fat droplets surround centrally located nuclei.

Swansea criteria is used, if six or more components are present in absence of other cause, it helps in diagnosis of AFLP [5]. It includes,

Table 1

Clinical	Vomiting
	Abdominal pain
	polydipsia/polyuria
	Encephalopathy
Biochemical-hepatic	Bilirubin >14µmol/l
	AST/ ALT > 42 IU/l
	Ammonia >47 µmol/l
Renal	Urate> 340µmol/l
	Creatinine >150µmol/l
Endocrine	Glucose < 4 mmol/l
Hematological	Leucocytosis >11*10^9/l
	Coagulopathy - PT > 14 secs or APTT > 34 secs
Radiological	Abdominal USS bright liver echo texture / ascites
Histological-Liver biopsy	Micro vesicular steatosis

The currently recommended mode of treatment of this catastrophic disease includes prompt delivery of fetus, regardless of gestation age, because delivery initiates the resolution of this life threatening disease.
It involves a multi-disciplinary team of Obstertrician, Hepatologist, Anaesthetist, Neonatologist in a tertiary care centre equipped with Intensive Care Unit [4].
The mode of delivery depends upon the current maternal condition and probability of a successful vaginal delivery, If vaginal delivery cannot be achieved quickly then Caesarian section is the preferred method.
The mortality rate of mothers who underwent LSCS (12.8%) was lower than who delivered vaginally (35.2%).
Mortality is attributed to the complications that can occur like Acute respiratory distress syndrome (ARDS), hepatic failure, hepatic encephalopathy, renal failure, nephrogenic diabetes insipidus, hypoglycemia, sepsis, DIC, hemorrhagic shock secondary to either intra-abdominal bleeding or upper gastrointestinal haemorrhage , culminating into multi organ dysfunction.
The maternal condition must be stabilised first in a ICU set up, if moderate to severe hepatic dysfunction is present. It involves supportive management with intravenous infusion, intravenous glucose, fresh frozen plasma and packed red blood cells if coagulation dysfunction is present, cryoprecipitate to correct DIC. Further course of management include the appropriate mode of delivery. Continuous close monitoring in the immediate post-partum period is equally very important. In most patients AFLP resolves almost completely after delivery with return of normal liver functions within 7 to 10 days.
Liver transplantation remains the last resort in fulminant

hepatic failure. Novel treatment modalities like Plasmapheresis, looks promising but still needs to be explored in detail [5]. The role of genetic testing also needs to be analysed. Suzuki *et al* , reported that the soluble fms-like tyrosinekinase 1/ placental growth factor ratio may be rapidly used to distinguish AFLP from HELLP syndrome.[6] More researches are to find way to predict AFLP.

Although the theoretical 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. If the patient decides to be pregnant again, she should be closely monitored for any early signs of acute fatty liver.

Conclusion

AFLP is uncommon, life threatening disorder with variable presentation. Hence a high index of suspicion is required for early diagnosis. It may occur rapidly and may progress unpredictably. Prompt delivery and intensive supportive care remain as the mainstay treatment for AFLP.

References

1. Hin Hin Ko, Eric Yoshida. Acute fatty liver of pregnancy. Can J Gastroenterol,2006:20(1):25-30.
2. Sheehan HL. The pathology of acute yellow atrophy and delayed chloroform poisoning.J Obstet Gynaecol Br Emp,1940:47:49-62.
3. Monga M, Katz AR. Acute fatty liver in the second trimester. Obstet Gynecol,1999:93:811-3.
4. Dwivedi S, Runmei M. Retrospective Study of Seven Cases with Acute Fatty Liver of Pregnancy , ISRN Obstet Gynecol,2013:2013:730569
5. Perera IA, Pradeepan JA, Mayorathan U *et al*. A case of fatal acute fatty liver of Pregnancy and Literature Review. Int J Reprod Contracept Obstet Gynecol,2018:7:2026-30.
6. Suzuki H, Nagayama S, Hiroshima C, *et al*. Markedly higher sFlt-1/PlGF ratio in a woman with acute fatty liver of pregnancy compared with HELLP syndrome. J Obstet Gynaecol Res,2019:45:96-103.
7. Ziki *et al*, Acute fatty liver of pregnancy. BMC pregnancy and childbirth,2019:19:259.