CASE REPORT

Acute fatty liver of pregnancy: Case report of an uncommon disease

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ABSTRACT

A 29 - year old female gravida 2 para 1 at 32 weeks 2 days of gestation presented with jaundice, itching, fever, vomiting, respiratory difficulty and altered sensorium. From clinical and initial laboratory findings diagnosis of acute fatty liver of pregnancy was made. Although early termination of pregnancy was done, postpartum period was complicated by sepsis, hepatic encephalopathy, pulmonary oedema, convulsion and continuing coagulopathy. Supportive management in an intensive care unit resulted in successful outcome.

Keywords: Acute fatty liver, pregnancy, jaundice, sepsis, DIC.
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Acute fatty liver of pregnancy (AFLP) is a serious complication among the various causes of pathological hepatic dysfunction in pregnancy. It is rare but potentially fatal complication for both mother and baby as often the diagnosis is delayed. Etiology is still unclear but known to be associated with defective fatty acid oxidation in foetus. AFLP is commoner in primigravida and there is an association with obesity, multiple pregnancies and male foetus (ratio 3:1). It is commoner in women aged 16-39 (mean age 29). The incidence is 1 in 7,000 to 1 in 15,000 pregnancies [1].

The mortality from acute fatty liver of pregnancy has been reduced significantly to 18%, and is now related primarily to complications, particularly DIC and infections [2, 3]. After delivery, most mothers do well, as the stimulus for fatty acid overload is removed. The disease can recur in future pregnancies, with a calculated genetic chance of 25%; the actual rate is lower, however [4]. Mortality of the foetus has also diminished significantly, but still remains 23% [5]. Indeed the recent UKOSS study which reported 61 cases showed no maternal mortality and 13% fetal mortality [6].

Case report

A 29 years old woman, gravida 2 para 1, currently with twin at 32 weeks 2 days of gestation was admitted to the hospital at emergency intensive care unit (ICU) with history of difficulty in respiration and altered sensorium for one day. There was also history of yellowish discolouration of the eyes, urine and body and also itching, malaise, nausea and vomiting since last twelve days. Supportive treatment for

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hepatitis was given at a private clinic.

Physical examination revealed a well-nourished, drowsy and icteric women. Her temperature was 100 degree F, pulse rate was 94/min, respiratory rate 26/min and blood pressure was 100/60 mm of Hg. Obstetrical examination revealed twin viable pregnancy. She was oriented to person, place and time.

Complete blood count revealed a haemoglobin: 13.5g/dl, white blood cell count 12,200/cumm and platelet count 1,40,000/cumm. Liver function test showed aspertate aminotransferase: 74 U/L, alanine aminotransferase 71 U/L, total bilirubin: 21mg/dl, direct bilirubin: 17mg/dl, alkaline phosphatise: 740 U/L, albumin: 2gm/dl. Biochemical test revealed blood urea: 40 mg/dl, serum creatinine 2.6mg/dl, random blood sugar 80mg/dl and serum ammonia: 76 mcg/dl. Coagulogram revealed a prothrombin time of 42 seconds, partial thromboplastin time 168 seconds, international normalized ratio (INR) of 3.23 and fibrin degradation product (FDP): 1678 ng/ml. Urine analysis showed no proteinurea. Serology test like HBsAg, HCV, HAV, HEV and HIV were all negative. Ultrasonography of the whole abdomen showed hyperechoic liver with ascites with viable twin intrauterine fetus. A presumptive diagnosis of AFLP was made.

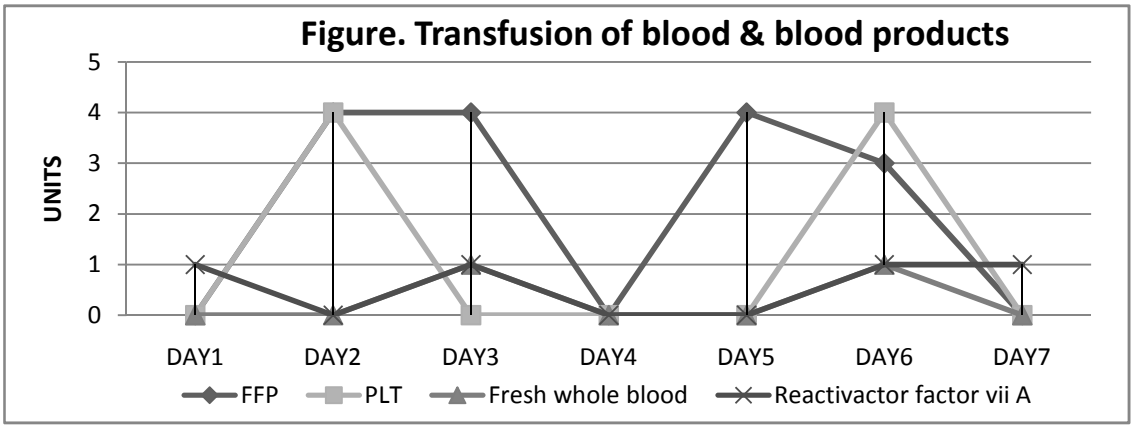
On consultation with the family members, it was decided to terminate the pregnancy. Misoprostol tablet was used to induce labour and she delivered two still born male child of 1.8kg and 2.2kg on second day of admission. Before which she received 10 mg vitamin K, 4 units of fresh frozen plasma, 4 units platelet, reactivation factor VII 1 mg to correct her coagulopathy and broad spectrum antibiotic. After delivery she was shifted to ICU again .On day 3 she had fever, tachycardia and blood pressure was 144/94mm of Hg with abdominal distension. On day 7, one episode of convulsion occurred and anticonvulsant drug started. On day 8, she developed pulmonary oedema with Spo_2 74% and was managed with 160mg of furosemide IV stat and nebulisation. On day 11 she had second episode of seizure. CT- Scan of brain showed bilateral parietal lobe white matter hypodensity. Complete blood count, liver function test, biochemical test and coagulation profile was fluctuating in abnormal range throughout the period in ICU. Other test reports were ANA negative, anti DsDNA negative, fibrinogen clotting activity 136 ng/dl, Coomb test - direct and indirect negative, laptospira antigen negative, peripheral smear was negative for haemolysis and malaria parasite, widal test insignificant, ascitic fluid analysis and culture normal, blood culture no growth, echocardiography normal and chest x-ray normal. During this period, she received fresh frozen plasma 15 units, platelet 8 units, fresh whole blood 2 units and reactivator factor VII 1 mg four doses. She made gradual recovery and was shifted from ICU to ward on day 20. Her liver and kidney function returned to normal and was discharged on day 36.

Table. Laboratory results

Investigation	DAY- 1	DAY- 3	DAY- 10	DAY- 13	DAY-21
Total bilirubin(mg/dl)	21	22	24	27	4.6
Tc (Cumm)	12,200	18,330	14,520	10,310	11,000
PLT (Cumm)	140,000	96,000	65,000	75,000	210,000
S. Creatinine (mg/dl)	2.6	2.9	0.9	2.6	0.35
AST (U/L)	74	67	53	-	103
ALT (U/L)	71	48	39	-	64
PT (Sec)	42	31	23.2	19.4	15
APTT (Sec)	164	62	52.8	47.5	-
INR	3.32	3.19	-	1.69	-

Discussion

Acute fatty liver of pregnancy is autosomal recessive in inheritance and mothers are often found to be heterozygous for the affected mutation [7]. The understanding of the causes of acute fatty liver of pregnancy has been ameliorat-ed by advancees in mitochondrial bio-chemistry. Deficien-cy of LCHAD (Long chain 3-hydroxyacyl-CoA dehydrogenase) leads to an accu-mulation of medium



and long chain fatty acids [8]. The accumulation of long-chain 3-hydroxyacyl metabolites produced by the foetus or placenta is toxic to the liver and may be the cause of the liver disease. Criteria for diagnosis of AFLP [9] include six or more of the following features in the absence of another explanation: Vomiting, abdominal pain, polydipsia / polyuria, encephalopathy, elevated bilirubin (>14 micro mol/l), hypoglycemia (<4 mmol/l), elevated urate (>340 micro mol/l) , leucocytosis (>11x10^9/l), ascites or bright liver on ultrasound scan, elevated transaminases (aspartate aminotransferase or alanine aminotransferase >42 IU/l), elevated ammonia (>47 micro mol/l), renal impairment (creatinine >150 micro mol/l), coagulopathy (prothrombin time >14 s or activated partial thromboplastin time >34 s), microvesicular steatosis on liver biopsy. The most striking feature of this syndrome is a high level of bilirubin associated with moderate increases of transaminases. Our patient had more than six of the above mentioned criteria. Initial treatment involves supportive management with intravenous fluids, intravenous glucose and blood products, including fresh frozen plasma and cryoprecipitate to correct DIC. Once the mother is stabilized, arrangements are usually made for delivery. This may occur vaginally, but, in cases of severe bleeding or compromise of the mother's status, a caesarian section may be needed [10].

Conclusion

AFLP is an uncommon, life threatening disorder developing in the third trimester of pregnancy or early postpartum period. Careful history and physical examination in conjunction with compatible laboratory and imaging results are often sufficient to make the diagnosis and liver biopsy is rarely indicated. Prompt delivery of the fetus and intensive supportive care remain the mainstay treatment for AFLP.

References

1. Gregory TL. Hughes S. Coleman MA. De Silva A. Acute fatty liver of pregnancy; three cases and discussion of analgesia and anesthesia. International Journal of Obstetric Anesthesia. 2007; 16 (2):1759.

2. Ko H, Yoshida EM. "Acute fatty liver of pregnancy". Can. J. Gastroenterol. 2006; 20 (1): 25–30.

3. Mjahed K, Charra B, Hamoudi D, Noun M, Barrou L. "Acute fatty liver of pregnancy". Arch. Gynecol. Obstet. 2006; 274 (6): 349–353.

4. Tein I. "Metabolic disease in the foetus predisposes to maternal hepatic complications of pregnancy". Pediatr. Res. 2000; 47 (1): 6–8.

5. Fesenmeier MF, Coppage KH, Lambers DS, Barton JR, Sibai BM. "Acute fatty liver of pregnancy in 3 tertiary care centers". Am. J. Obstet. Gynecol. 2005; 192 (5): 1416–1419.

6. Rajasri AG. Srestha R. Mitchell J. Acute fatty liver of pregnancy (AFLP) - an overview. Journal of Obstetrics & Gynaecology. 2007; 27(3): 237-40.

7. Wanders RJ, Vreken P, Den Boer ME, Wijburg FA, van Gennip AH. Disorders of mitochondrial fatty acyl-CoA beta-oxidation. J Inherit Metab Dis. 1999; 22(4): 442-87.

8. Tein I. Metabolic disease in the fetus predisposes to maternal hepatic complications of pregnancy. Pediatr Res. 2000; 47(1): 6-8.

9. Ch'ng CL, Morgan M, Hainsworth I, et al. Prospective study of liver dysfunction in pregnancy in Southwest Wales. Gut. 2002; 51: 876–80.

10. Ko HH, Yoshida EM. Acute fatty liver of pregnancy. Can J Gastroenterology. 2006; 20(1): 25-30.