

A Case of Acute Fatty Liver of Pregnancy

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We described a case of acute fatty liver developed early postpartum period. Acute fatty liver of pregnancy (AFLP) is a rare medical condition and is important due to its implications on maternal and fetal morbidity and mortality. AFLP is managed with supportive care and close surveillance is needed in these patients. We report the case of a previously healthy 29 year old woman within the postpartum first day who presented to the emergency department with postpartum severe vaginal bleeding. An evaluation revealed hepatic and renal failure. The patient required aggressive supportive care, transfusion of multiple blood products in intensive care unit. This case was unusual because AFLP was reported in postpartum period and the patient presented due to postpartum severe vaginal bleeding after vaginal delivery and was subsequently diagnosed as AFLP

Key Words: Acute fatty liver, Pregnancy

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Case Report

29 year-old woman admitted to emergency room in the first day of postpartum with massive vaginal bleeding after normal spontaneous vaginal delivery. She had previous three pregnancies delivered vaginally without any complication. Physical examination revealed a well developed young woman with blood pressure 90/ 60 mmHg, heart rate 110 beats/min, respiratory rate 25 breaths /min and temperature 37 °C. The abdomen was non-tender without hepatomegaly. The uterus was found to be above the level of umbilicus with moderate vaginal bleeding. Neurologically, the patient was conscious and oriented. Full blood count and liver function tests were performed at the time. Hemoglobin was 8.7 mg/dl. The patient had hyperbilirubinemia with direct and indirect bilirubin increased to 10. 6 mg/dL and 8.26 mg/dL respectively; aspartate amino transferase enzyme (AST) was 57 U/L (normal range 8-46 U/L), alanine aminotransferase enzyme (ALT) was 53 U/L (normal range 7-46 U/L) and glucose was 112 mg/dl. Coagulation tests were also abnormal with prolonged prothrombin time (PT) >120 sec (normal range 10-14 sec) and partial thromboplastin time (APTT) > 190 sec (normal range 22-35 sec). White blood cell was 12.2 10³ /uL (normal range was 3.58-11.07 10³ / uL), thrombocytes was

185000 / mm³ (normal range 165.4-352.9 10³ /uL). A virology screen performed for Hepatitis A, B, C, E viruses; cytomegalovirus; and Epstein Barr virus were all negative. As the patient's clinical situation went worse and her Glasgow Score (GCS) was decreased to ten following two days, she was transferred to the intensive care unit (ICU). As the patient had unconscious and had respiratory difficulty and a productive cough invasive monitoring was started. A Computerizing Tomography (CT) scan was performed, but there was no evidence of intracranial hemorrhage or post-ischemic changes. In arterial blood gas analysis was shown that there was significant metabolic acidosis with pH;7.15; pCO₂;30; pO₂ ;135,6; BE(B);-16.9; BE (ecf);-18.5; and temperature: 36.0 °C, Hb: 7.4 gr/dl, WBC: 16.5 10³/uL, Platelets: 138 10³/uL, so mechanical ventilation was initiated in sixth day of ICU. The picture of metabolic acidosis continued and renal impairment with reduced urine output was developed. Ultrasound scans of the upper abdomen revealed basal bilateral pleural effusion and with normal liver architecture. CT scan of the upper abdomen revealed that there was edema in periportal area and slightly fatty appearance of the liver, pancreas and gall bladder were normal. After careful research of clinical and laboratory findings of the patient, the diagnosis of acute fatty liver of pregnancy was made and supportive management was continued. Following days, her condition was deteriorated and her serum liver function tests increased progressively. At 5th day, the liver function tests showed increased AST of 144 U/L and ALT of 47 U/L. At 10th days, serum ammonium, Blood Urea Nitrogen (BUN) and creatinine levels were increased to 138.3 mg/dL, 6.11 mg/dL, and 61.35µg/dL respectively. Blood glucose decreased to 36 mg/dl. Serum direct and indirect bilirubin increased to 30.35 mg/dL and 22.66 mg/dL respectively; At the 15th day of admission to intensive care unit, the

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patients' laboratory tests were started to return to normal ranges gradually. Clinical condition of the patient became stable and the patient weaning procedure was started with easy-breath, however the patient consciousness was not normal, her GCS was still ten. Her clinic was going to better day by day. She recovered gradually with rehabilitation and physiotherapy and discharged from the hospital 23 days later after admission to ICU. At a recent follow up she was good without any major sequelae.

Discussion

Acute fatty liver of pregnancy (AFLP) is a rare condition which is seen about 1/7000- 1/ 16.000 pregnancies.¹ It was first identified by Sheehan in 1940.² While the etiology of AFLP is unclear; a genetic component has been suggested. AFLP is also thought to be associated with gene mutation in the long chain 3-hydroxyacyl-coenzyme a dehydrogenase, a fatty acid β oxidation enzyme.³ Maternal and fetal mortality rates are estimated to be 18% and 23% respectively. It almost always occurs in the third trimester of gestation. Very occasionally, it has been diagnosed in the first few days postpartum as in the present case.

The commonest clinical presentation is pain, jaundice and vomiting with features of encephalopathy. After delivery, most patients improve slowly and a full clinical and laboratory recovery may take 1-6 weeks. The abnormal laboratory parameters are conjugated hyperbilirubinemia, increased alkaline phosphates and moderate elevation of transaminases. AFLP is suspected by jaundice, liver failure, and hyperammonemia.⁴ Leucocytosis occurs commonly with thrombocytopenia and abnormal clotting. Common complications associated with AFLP are cerebral edema, renal failure, hypoglycemia, infections, gastrointestinal hemorrhage, coagulopathy and fetal death. Abnormal liver tests occur in pregnancies with many potential causes, including viral hepatitis, preeclampsia, HELLP syndrome (Hemolysis, Elevated Liver Enzymes and Low Platelets) and intrahepatic cholestasis. Hepatitis A, B and C infections can cause high liver enzymes, coagulopathy and hyperbilirubinemia. In the presented case, virology screens were negative. In contrast to viral and other fulminant hepatitis there is relatively minor necrosis of hepatocytes in AFLP.⁵

Sherlock et al showed that there are swelling of the lobules, compression of the sinusoids and micro vesicular, centrilobular fatty infiltration of hepatocytes in the liver biopsy of patient with AFLP.⁶ Therapeutic decisions were made without waiting for a confirmed histological diagnosis. The liver biopsy should not be performed routinely to differentiate the AFLP from severe preeclampsia. Because both supportive care and delivery is the mainstay of management for both diseases.⁷ Tendency of the high risks of bleeding also restrict the clinicians to perform liver biopsy. CT scan assisted our clinical

diagnose without need liver biopsy.

Although ultrasound is important diagnostic tool in exclusion of biliary tract disorders its value in AFLP is limited like CT and MR (8). Clinical trials showed that there is no specific treatment for this condition however, hospitalization and supportive care is the mainstay of management. There are no published reports of recovery before delivery and hence a prompt delivery is advisable (9). Patients who have experienced AFLP should be warned about the risk of recurrence in subsequent pregnancies.

AFLP is an obstetric and medical emergency because of the associated metabolic changes and complications and may need to terminate pregnancy. Treatment of AFLP is largely supportive and best carried out in a critical care environment with multidisciplinary input. Close follow up of future pregnancies in patient affected previously with this disease is recommended.

Gebeliğin Akut Yağlı Karaciğeri Olgu Sunumu

Olgumuz, erken postpartum dönemde karaciğerin yağlı nekrozu tanısı alan bir lohusadır. Gebelikte görülen karaciğerin yağlı nekrozu, maternal ve fetal morbidite ve mortaliteye sebep olması nedeniyle nadir görülmekle beraber, önemli bir gebelik hastalığıdır ve tedavisinde yakın gözetim ve destek tedavisi önemli rol oynar. Hastamız, gebeliğinde herhangi bir sorunu olmayan, postpartum birinci gününde ciddi vajinal kanama ile acil servisimize başvuran 29 yaşında bir lohusa idi. Yapılan değerlendirme sonucu, hastada hepatic ve renal yetmezlik tespit edildi. Hasta yoğun bir destek tedavisine ve çok kez kan ürünleri transfüzyonuna ihtiyaç duydu. Olgumuz gebelikte görülen karaciğerin yağlı nekrozunun postpartum dönemde ortaya çıkması ve ilk bulgunun şiddetli vajinal kanama olması ve sonrasında tanının konulması ile farklı bir olgudur.

Anahtar Kelimeler: Akut yağlı karaciğer, Gebelik

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