

Acute Fatty Liver of Pregnancy

Dear Editor,

Acute fatty liver of pregnancy (AFLP) is a rare, potentially life-threatening, pregnancy-related disease that affects 1 in 7000 to 16,000 pregnancies.^[1] The condition occurs more commonly in primigravidas, multiple pregnancy, and pregnancies carrying a male fetus.^[2] At presentation, one should keep in mind other pregnancy-related liver disease that mimic AFLP such as HELLP syndrome (hemolysis, elevated liver enzymes, and low platelet count). Bleeding and disseminated intravascular coagulation (DIC) are one of most common complications.

We report that a 21-year-old primigravida (BMI - 20.4 kg/m²) with twin pregnancy (diamniotic-dichorionic), admitted with labor pains at 35 weeks and 3 days period of gestation with blood pressure of 150/96 mm Hg and urine albumin of 1+. Her biochemical and hematological parameters were within normal limits. She delivered 2 live female babies by vaginal delivery with episiotomy weighing 2.25 kg and 1.9 kg at 20:25 hrs and 20:28 hrs, respectively. Post-delivery at 02:00 hrs, vulval hematoma was drained in OT. Postoperatively, she was found to have pallor and minimal vaginal oozing. She was given 2 PRBC and 10 mg vitamin K. Four hours later, blood clots were found in vagina (approx 800 ml). Exploration under GA was done. Tear was excluded. There was active bleeding from the uterus, oozing from the episiotomy site. Vaginal packing, inj. PGF2 alpha 02 doses at 20 min apart, 4 units of PRBC and 8 FFP were given in the OT, and she was shifted to ICU. Hematological investigations were suggestive of consumptive coagulopathy with deranged liver enzymes and coagulation parameters [SGOT: 756 IU/L, SGPT: 356 IU/L, LDH: 2754 IU/L, INR: 3.19, platelets 130,000/mm³, PT: 32 sec (13), PTTK: 39 sec (27), TLC 19,000/mm³]. She was given injectable antibiotics, FFP 2 units 6 hourly and cryoprecipitates 8 units. She was put on ventilator at 15:00 hrs in P-SIMV mode, PEEP 5, and respiratory rate 15/min. Investigations 6 hours later showed increasing liver enzymes, INR 2.94, platelets 48,000/mm³, Hb 6.5 gm% with low urine output. In view of ARF and risk of fluid overload, dialysis was started along with frusemide infusion. She continued on blood products. USG abdomen revealed hepatomegaly with fatty changes grade 2-3, medical renal disease, and minimal ascites. Vaginal packing removed after 24

hours and oozing was still present. Repacking was done. Liver enzymes kept on increasing (maximum serum bilirubin 15.5 mg%, SGOT 3450 IU/L, SGPT 2600 IU/L on postpartum day 3) and clinically she was in grade 1 to grade 2 encephalopathy. Viral markers for hepatitis A, B, C and E were negative. Following hemodialysis for the fourth continuous day, her vitals were found to be stabilized. Vaginal pack was removed after 48 hours of repacking and she was extubated on fourth postpartum day. LFT though decreasing continued to be deranged. Urine output was 450 - 500ml/24 h. Hemodialysis was done every alternate day till 16th postpartum day. During this period she developed ascites and bilateral pleural effusion owing to hypoproteinemia for which she was treated with albumin. Alternate day USG carried out and showed improvement in fatty changes, disappearance of ascites and pleural effusion. Liver enzymes normalized on 19th postpartum day. Urine output started increasing and 25th postpartum day she was discharged.

Acute fatty liver almost always manifests late in pregnancy with a mean gestational age of 37.5 weeks;^[3] some do not become clinically evident until delivery.^[4] Persistent nausea and vomiting are major symptoms with half of affected women might have hypertension, proteinuria and edema, alone or in combination. According to the Swansea criteria,^[5] six or more of the following features are used to diagnose AFLP in the absence of other explanations: vomiting; abdominal pain; polydipsia/polyuria; encephalopathy; elevated bilirubin >14 µmol/L (0.8 mg%); hypoglycemia <4 mmol/L (72 mg%); elevated urate >340 µmol/L (5.7 mg%); leukocytosis >11 × 10⁹/L; ascites or bright liver on ultrasound; elevated transaminases; elevated ammonia >47 µmol/L (27.5 mg%); renal impairment creatinine >150 µmol/L (1.7mg%); coagulopathy (PT >14 sec or APTT >34 sec), or microvesicular steatosis on liver biopsy. Using these criteria, we anticipated the diagnosis of AFLP. The patient had elevated bilirubin, elevated transaminases, elevated creatinine, encephalopathy, leukocytosis, fatty liver grade 2-3 and coagulopathy. A patient with severe preeclampsia/HELLP will usually present with proteinuria.

Systemic complications of AFLP are due to fulminant hepatic failure and include encephalopathy, acute renal failure, infection, pancreatitis, gastrointestinal hemorrhage, coagulopathy, and at least mild hypoglycemia. Symptoms may rapidly progress from restlessness, confusion, and disorientation to asterixis, seizures, psychosis, and ultimately coma.^[6] Other systemic effects include respiratory failure, sometimes requiring assisted ventilation, ascites,^[4] and gastrointestinal bleeding

from gastric ulceration and Mallory-Weiss syndrome.^[7] Hepatorenal syndrome eventually develops and leads to oliguria and acute tubular necrosis.^[6] Ultrasound and CT scans of the liver have been used for diagnosis, but the specificity and sensitivity of these studies are insufficient to make a diagnosis and the likelihood of false negative results is high.^[4] Liver biopsy is the gold standard test, but it is invasive and requires a patient with normal coagulation status.^[3]

Supportive care of patients with AFLP should include careful monitoring for evidence of progressive hepatic failure, hypoglycemia, and coagulopathy. This should occur in an intensive care setting and in consultation with physicians well-versed in the care of critically ill patients. Spontaneous resolution usually follows delivery. Maternal deaths are caused by sepsis, hemorrhage, aspiration, renal failure, pancreatitis and gastrointestinal bleeding.^[8] Although maternal mortality rates in the past approached 75 percent but Sibai (2007) cites an average mortality rate of 7 percent with 70 percent preterm delivery rate and perinatal mortality rate of approximately 15 percent, which in the past was nearly 90 percent.^[9] Early diagnosis, prompt therapy, adequate supportive care and a multidisciplinary approach are the key to a good outcome.

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