

ACUTE FATTY LIVER OF PREGNANCY-A CASE REPORT

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Background

Acute fatty liver of pregnancy (AFLP) is a rare but potentially lethal disease affecting both the mother and the foetus. It is usually a diagnosis of exclusion, requiring a strong index of suspicion, and is more common in primigravidae. There is an association with obesity, male foetuses, and multiple pregnancies.

We present a case report of a 22-year-old woman with AFLP-unrecognised, complicated by stillbirth, DIC, and severe hypoglycaemia, who required transfer to the intensive care unit (ICU) at a Liver Unit.

Case Report

A 22-year-old woman G2 P1 at 33 weeks +3 days, was admitted to the hospital after having a pre-term stillbirth male baby at home. She was then transferred to the hospital.

She reported that she had reduced foetal moments, polydipsia, and polyphagia for 2 days, and epigastric pain, nausea, and vomiting for one week prior to her delivery.

She had tachycardia on arrival, icteric with bilateral pedal oedema, her abdomen was non-tender with a contracted uterus and moderate vaginal bleeding.

A complete blood picture revealed a haemoglobin of 150 g/dL, white blood cell 22.4x10<sup>3</sup>/L, platelets of 166×10<sup>9</sup>/L, bilirubin 204 mg/dL, ALT 557 U/L, ALP 557 U/L, profound hypoglycaemia <1.1 mg/dL, activated partial thromboplastin time (APTT) -35.7, APTT -96.2, and fibrinogen of 0.5

Within a couple of hours of arriving at the hospital, she became more icteric and appeared confused.

The lab picture fit with the Swansea criteria and along with the clinical presentation, a presumptive diagnosis of AFLP was made.

The patient was given 20% glucose intravenously and blood glucose was maintained at around 6 mg/dL; two units of cryoprecipitate were given because of the impending DIC. She was then transferred to the Liver Transplant Unit at Kings College Hospital for further management and intensive care. She underwent an examination under anaesthesia and evacuation of retained products of conception. She was started on iv antibiotics cefuroxime and metronidazole and low-molecular-weight heparin for VTE prophylaxis. Her renal function significantly improved with supportive care. She made a gradual recovery and her blood picture returned to normal at her follow-up. The patient refused a post-mortem examination of the baby and any other genetic testing including long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency screening.

Discussion

AFLP is rare with an incidence of 1 in 9000 to 1 in 13,000 pregnancies. Hypoglycaemia, jaundice, and coagulopathy with marked elevation of transaminases are the key features.

LCHAD deficiency is a recently described disorder of mitochondrial fatty acid oxidation that is usually asymptomatic in heterozygous carriers. Affected individuals often die in early childhood or sooner due to complications of hypoglycaemia, cardiomyopathy, or fatty liver failure.

One in 62,000 pregnancies results in LCHAD deficiency. Affected foetuses do not metabolise long-chain fatty acids completely, resulting in the accumulation of abnormal and highly toxic intermediates in the mother’s liver and the acute clinical picture that we see in AFLP. It is also suggested that fat accumulation may occur in the placenta and that this may induce the clinical picture of pre-eclampsia.

An individualised decision regarding the severity of maternal and foetal conditions needs to be made. Often induction of labour can be started while the maternal condition is stabilized, and blood products made available.

Initial treatment involves supportive management with glucose and blood products including FFPs and cryoprecipitate for coagulopathy. One-to-one care in the delivery suite is required, preferably in a high-dependency unit. Observations should be monitored and clearly documented on a 24-hour spreadsheet along with glucose monitoring at least every two hours. PT should be monitored every 6 hours along with LFT, FBC, urea, and electrolytes. A formal assessment of consciousness levels using the Glasgow Coma Scale should be made hourly because coma is a potential complication of AFLP. One of the keys to successful management is a multidisciplinary approach. Senior obstetricians, anaesthesiologists, and hepatologists should be involved at an early stage. This will also facilitate and alert the transplant team

FOLLOW-UP. All women affected by ALFP should have the opportunity with their partners to be debriefed about the complications of their pregnancy. Consideration of screening all babies of mothers with AFLP for LCHAD deficiency should be made. The recurrence rate is 15-25% for future pregnancies with the same partner if the baby is positive for LCHAD deficiency. CVS/amniocentesis is also another option in subsequent pregnancies.

The definitive treatment is rapid delivery and supportive care, which reduces mortality. Liver transplantation is rarely required but may be needed for mothers with severe DIC.

