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A Case of Massive Hepatic Necrosis in a Patient with AFLP; An Interesting Case of Obstetric Emergency

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Abstract: ***Background: Acute fatty liver in pregnancy (AFLP) is an obstetric emergency and carries high morbidity and mortality, including death in the mother and her fetus. It is characterized by maternal liver failure. Maternal mortality has decreased to 4% in recent years due to quicker diagnosis and immediate delivery. AFLP is not a predictable or preventable condition. Live failure, coagulopathy, hemorrhage, renal failure, and severe infection can be lethal for both the mother and fetus. Case report: Patient is a 30 yr old female referred from as a case of preterm labour admitted as Primigravida at 33.5wk (d) with preterm labour. USG was done and patient taken for emergency LSCS I/v/o suspected abruption. Postoperative patient had one episode of GTCS hypoglycemic seizure investigations revealed raised SGOT/SGPT. I/V/O breathlessness patient shifted to ICU intubated. Patient went into AKI started on dialysis went in DIC coagulation failure transfused accordingly. Patient expired on 7th POD. Baby discharged.***

Keywords: AFLP, Hepatic necrosis, Liver, Preterm

1. Background

Acute fatty liver in pregnancy (AFLP) is an obstetric emergency and carries high morbidity and mortality, including death in the mother and her fetus. Acute fatty liver of pregnancy is an uncommon but potentially fatal complication that occurs in the third trimester or early postpartum period. It was first described in 1940 by Sheehan as an “acute yellow atrophy of the liver”. It is characterized by maternal liver failure. Maternal mortality has decreased to 4% in recent years due to quicker diagnosis and immediate delivery. AFLP is not a predictable or preventable condition. Liver failure, coagulopathy, hemorrhage, renal failure, and severe infection can be lethal for both the mother and fetus. Pregnancy - related liver disease includes several distinct liver disorders that affect different stages of pregnancy; these include acute fatty liver of pregnancy, pre - eclampsia liver dysfunction, and hemolysis, elevated liver enzymes, and low platelets syndrome (HELLP).

The incidence of AFLP was initially reported to be very low, between 0.001% and 0.015%, ([4, 5]) with a high maternal and neonatal mortality rate of up to 70%. . It usually presents in late pregnancy and can result in maternal or fetal complications. It is characterized by acute liver dysfunction due to fatty infiltration of liver parenchyma, which can precipitate coagulopathy, electrolyte imbalance, and multi - organ failure. Early recognition and treatment of this disease entity are essential as it carries high morbidity and mortality. Along with supportive care for the mother, delivery of the fetus is the only definitive treatment of this condition.

2. Case Report

Patient is a 30 yr old female referred as a case of preterm labour admitted in the hospital as Primigravida at 33.6wk (d) with preterm labour. Patient taken for emergency LSCS in view of persistent fetal tachycardia with suspected abruption. There were no significant OT findings were normal except 150cc of straw - colored ascitic fluid.0n first post op day pt had an episode of hypoglycemic seizure Investigations were repeated following this, which showed thrombocytopenia (35, 000), increased bilirubin (T/D - 2.7/1.26), OT/PT (351/128). In view of thrombocytopenia, the patient transfused 4 pints of platelets and started on Inj vit K and ursodeoxycholic acid. CT brain was normal. The repeat platelet count was 60, 000 transfused 4 - pint platelets. On the second POD, the patient had breathlessness and was shifted to the ICU. Investigations revealed Hb - 7.7 g/dl platelet count 67, 000, S. Creatinine 1.23mg/dl. Further derangement of LFT S. bilirubin (T/D - 5.3/2.7) SGOT/SGPT (4202/2386) T. Protein - 4.6gm INR - no coagulation s. LDH (9935) ABG showed acidosis, in view of the above patient transfused one pint of packed cells, four pints of platelets, four pints of FFP, and eight pints of cryoprecipitate. Patient intubated and ventilated on PRVC mode. USG is suggestive of fulminant hepatitis? infarction. In view of rising creatinine and reduced urine output, the patient underwent hemodialysis. On the 3rd POD, investigations were repeated, which showed Hb (6.8 g/dl), platelet count (60, 000), and INR - no coagulation. in view of the above patient transfused 1 pint packed cell, 4 pints cryoprecipitate, 4 pints FFP, and 4 pints platelets. Repeat investigations showed worsening of renal and liver function tests, with the INR showing no coagulation and nil urine output.2Decho showed severe TR and moderate MR. In view of hypotension, the patient was started on inotropes. The patient had a cardiorespiratory arrest on the 7th POD and expired. Baby discharged.

Liver biopsy done - HPE gross - brownish white elongated bits measuring 1.5cm. Microscopy - sections shows massive necrosis of hepatocytes and areas of lymphocytic infiltrate.

3. Discussion

Acute fatty liver of pregnancy sometimes presents as a diagnostic challenge, as it may be challenging to differentiate from HELLP syndrome and pre - eclampsia

with severe features. There is a significant overlap between these disease entities, and in fact, they may co - exist in the same patient. Signs of liver failure such as coagulopathy and disseminated intravascular coagulation and hypoglycemia are more consistent with AFLP as compared to HELLP or pre - eclampsia. The exact cause of AFLP is still unknown, however, it has been linked to mitochondrial dysfunction in fatty acid oxidation processes. Additionally, a deficiency in long - chain fattyacylhydroxy - CoA dehydrogenase (LCHAD), which is an autosomal recessive disorder has also been associated with AFLP. [(2)] In fact, pregnancies with LCHAD - deficient fetuses are more likely to develop AFLP, with up to 79% of these pregnancies affected. [(6, 7)]

In cases of fetal LCHAD deficiency, there is an accumulation of 3 - hydroxyfatty acids in the placenta, including 3 - hydroxymyristic acid, 3 - hydroxypalmitic acid, and 3 - hydroxydicarboxylic acid, as the fetal part of the placenta has the same genetic makeup as the fetus. When maternal hepatocytes are exposed to high levels of free fatty acids, the mitochondrial volume can become overwhelmed, leading to oxidative shunting of fatty acids into peroxisomes and oxidative stress in these organelles. [(1)] This can result in mitochondrial dysfunction and liver exposure to oxidative emergencies, which can lead to inflammation and fibrosis, ([(8)] ultimately inducing severe acute maternal liver failure. [3] Transaminase levels and hyperbilirubinemia is also more severe in AFLP. The Swansea criteria [(13, 14, 15)] are currently the internationally accepted diagnostic criteria for AFLP. These criteria have been found to have a sensitivity of 100%, specificity of 57%, positive predictive value of 85%, and negative predictive value of 100%. Diagnosis of AFLP requires the presence of six or more criteria. The severity of AFLP and the intensity of therapeutic intervention are determined by the score obtained using these criteria. It is generally believed that early diagnosis and differential diagnosis can be achieved without liver biopsy by combining clinical manifestations with laboratory tests. Early and accurate diagnosis, timely termination of pregnancy, and multidisciplinary supportive care are crucial in the management of patients with AFLP. [(9, 10, 11)] Both domestic and foreign guidelines for the treatment of liver failure recommend prompt termination of pregnancy in patients with AFLP, with delivery initiated within 24 h of diagnosis. The mode of delivery is determined based on maternal and fetal condition, gestational age, fetal position, and the likelihood of successful induction of labor. If a rapid delivery is not feasible, a cesarean section is necessary. In recent years, cesarean delivery has been recommended to improve fetal outcomes, as it has been shown to be the safest mode of delivery. Wang et al reported a 48% reduction in maternal mortality and a 44% lower perinatal mortality rate with cesarean section compared to vaginal delivery. [(9)] The duration of recovery for a woman after delivery is contingent on the severity of the disease and the presence of any additional complications. Most patients achieve clinical recovery within three to four days of delivery, although normalization of laboratory tests may be delayed and persist for days or weeks following delivery. Artificial liver support therapy can also be administered for women with persistent liver failure, which can improve liver damage and create a homeostatic environment for hepa tocyte regeneration. Various artificial liver supportive therapies, including hemoperfusion, plasma exchange (PE), continuous renal replacement therapy, molecular adsorption recirculation, and plasma perfusion (PP), are available. If a patient is experiencing end - stage liver failure, liver transplantation should be considered. [(16)] Plasmapheresis in theory can lead to the removal of ammonia, endotoxins, bilirubin, and inflammatory cytokines from the circulation that must be performed by liver cells. Also, injection of large volumes of FFP in this method can help to improve the DIC and removing renin angiotensin and other vasoactive factors may improve renal function. All these advantages improve hepatic and renal function in patients with AFLP. Therefore, this treatment especially considering the advanced cases of AFLP is very important.

4. Conclusion

AFLP is a rare, life - threatening complication of third trimester which requires a high index of suspicion for early diagnosis. Urgent delivery and maximum supportive care should be instituted to prevent poor outcome.

Conflict of interest: none

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