

Management of Rare Case of Acute Fatty Liver of Pregnancy with Pituitary Apoplexy and Disseminated Intravascular Coagulation Resulting in Foetus Death and Hemorrhagic Shock: A Case Report

Imane Laghrich^{1*}, Soukaina Laaraj¹, L Abdellah Babahabib¹, Moulay El Mehdi Elhassani¹, Jaouad Kouach¹

¹Gynecology Obstetrics Department, Mohamed V Military Training Hospital, Rabat, Morocco

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*Corresponding author: Imane Laghrich

Gynecology Obstetrics Department, Mohamed V Military Training Hospital, Rabat, Morocco

Abstract

We present a case of a 26-year-old female at 37 weeks of gestation with concomitant pituitary apoplexy and acute fatty liver complicated with disseminated intravascular coagulation, leading to foetus death and hemorrhagic shock secondary to a massive uterine haemorrhage which was successfully managed in Intensive Care Unit. To the best of our knowledge, this is the fourth case reported in the literature.

Keywords: Pituitary apoplexy; Sheehan syndrome; acute fatty liver; disseminated intravascular coagulation.

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INTRODUCTION

In 1937 Sheehan first published a report describing pituitary necrosis at the time of autopsy in women who died from obstetric hemorrhage [1]. Due to improvement of knowledge in medical care, specifically blood product and fluid replacement, the number of women surviving profound postpartum hemorrhage has increased [2]. The presence of disseminated intravascular coagulation (DIC) in the setting of acute fatty liver of pregnancy (AFLP), as well as blood loss are associated with labor increase risk of bleeding or ischemic necrosis, precipitating the pituitary apoplexy [3].

CASE REPORT

A written informed consent was obtained from the patient to publish this observation. A 26-year-old female who had previously given birth to a healthy child by normal delivery, without any relevant history, was admitted to the department of Gynecology obstetrics at 37 weeks of gestation due to the appearance of moderate jaundice complicated within few days with reduced frequency of fetal movements.

The physical examination revealed a conscious jaundiced female with icteric sclerae, hypertension and proteinuria. Oedema and neurosensory signs were not

found, and there was no hepatomegaly. Fetal heart rate was not noted. Pelvic ultrasound revealed an intrauterine pregnancy, biometrics corresponded to gestational age and a normal amniotic fluid index. No intra or retroplacental hematomas were noticed.

Her laboratory analysis showed elevated liver enzymes (Aspartate aminotransferase ASAT and Alanine amino transferase ALAT were 10 times above the reference value), decreased thrombin time at 38%, hemolysis with haemoglobin at 11g/dl, coagulopathy (low prothrombin time, decrease in fibrinogen and increased level of Ddimer), elevated creatinine at 21mg/dl and proteinuria at 0,8g/24h. No low platelet count was noticed. An abdominal ultrasound showed increased echogenicity of the liver (brightness) suggesting fatty infiltration without any evidence of intra or extrahepatic biliary dilatation.

The patient underwent induction and the fetus was delivered vaginally. A massive postpartum uterine haemorrhage was reported despite preventive measures, which was managed medically within few hours after delivery. The patient's haemoglobin level decreased from 7.9 to 5.7 mg/dl with low platelet account at 90000 and low fibrinogen suggestive of DIC. The

worsening of renal function with oliguria had also been noticed.

The situation has been controlled in the intensive care unit, using a massive blood transfusion including packed red blood cells, platelets and plasma. The urine flow was restored after fluid resuscitation and transfusion. Thereafter, the patient reported headaches and confusion, as well as significant decrease in breast milk.

Her laboratory tests were significant for persistently hypoglycaemia and low serum sodium

levels, what prompted a hormonal workup; Serum cortisol in the early morning was low, Follicle stimulating hormone (FSH), Luteinizing hormone (LH), Thyroid stimulating hormone (TSH) and prolactin were also low. These results supported the diagnosis of panhypopituitarism.

Magnetic Resonance Image scanning of the brain revealed ischemic lesions in the pituitary gland (necrosis) suggesting that the pituitary dysfunction was due to Sheehan's syndrome (Figure 1).

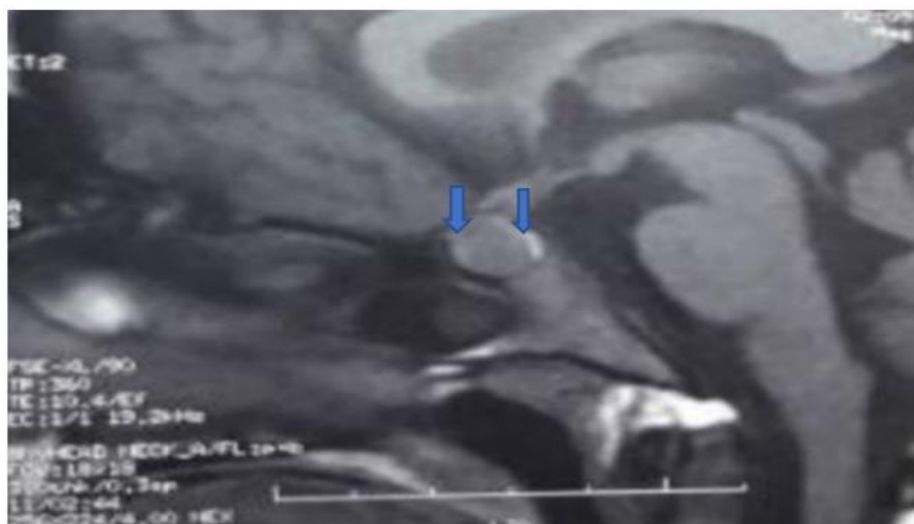


Figure 1: Sagittal MRI showing ischemic lesions in pituitary gland

During the follow up, serum liver enzyme levels gradually decreased with normalisation of renal function and improvement of hemostasis test, apart from persistent anemia related to hemolysis. Glucocorticoid and thyroid hormone replacement therapies were initiated with close follow-up in the endocrinology department.

DISCUSSION

We present a case of pituitary apoplexy consecutive to AFLP. To our knowledge, this is the fourth case reported in the literature (only four cases have been reported in literature so far) The pituitary gland is one of the most affected organs with altered anatomy and physiology during pregnancy. It is enlarged as a result of lactotroph hyperplasia [4].

Here, the blood supply to the already enlarged pituitary gland is seriously compromised in times of acute volume depletion compounded by vasospasm due to circulating vasoconstrictors. The enlarged gland and low pressure in the portal system cause susceptibility to tissue hypoperfusion and infarction. Thus, acute blood loss, acute stress, and coagulopathy secondary to acute liver failure may increase the risk of pituitary apoplexy [3].

The role of autoimmunity in the development of hypopituitarism has been suggested [5]. The disease often runs a prolonged course, with symptoms of pituitary insufficiency appearing years or even decades after the index delivery. It is rarely diagnosed in the acute peripartum period. Our patient had symptoms only few days after delivery [6].

Acute fatty liver of pregnancy (AFLP) is a maternal liver disease unique to pregnancy. The pathogenic mechanism of AFLP is a mitochondrial dysfunction causing defect in fatty acid. As the energy demands amplify in late pregnancy, a compensated defective fatty acid oxydation becomes overt as a result of increased reliance on fats as an energy source during late pregnancy [7]. Thus, intermediate products of metabolism can accumulate in maternal blood and hepatocytes, with deleterious effects on maternal hepatocytes.

The diagnosis of AFLP can be challenging because the initial clinical presentation is not always specific. The patient's history, clinical features and laboratory abnormalities may simulate conditions such as acute viral hepatitis, pre-eclampsia, HELLP syndrome, intrahepatic cholestasis or others [8]. As AFLP is uncommon, the best approach in case of liver

dysfunction during pregnancy is to rule out other causes.

In our case, DIC induced by AFLP and hemorrhagic shock, caused ischemia in the pituitary gland leading to Sheehan syndrome. Initial management of the patient with AFLP includes prompt delivery, regardless of gestational age. Intensive monitoring and prolonged supportive management including plasma exchange is required. Hypopituitarism is commonly treated with supportive measures, fluid management, electrolyte monitoring, and replacement of the deficient pituitary hormones [7].

CONCLUSION

The coexisting of AFLP and Sheehan's syndrome is rare. Both diseases could be related to the hypercoagulable state and uterine haemorrhage secondary to DIC. Prompt delivery as well as prompt initiation of hormone replacement therapy is essential to reduce morbidity and mortality risk.

Conflicts of Interest: Authors do not declare any conflicts of interest.

REFERENCES

1. Stander, H. J., & Cadden, J. F. (1934). Acute yellow atrophy of the liver in pregnancy. *Am J Obstet Gynecol*, 28, 61-69.
2. Sheehan, H. L. (1940). The pathology of acute yellow atrophy and delayed chloroform poisoning. *BJOG: An International Journal of Obstetrics & Gynaecology*, 47(1), 49-62.
3. Barvalia, U., Eliades, M., & Melhem, L. (2015). Concomitant Pituitary Apoplexy and Acute Fatty Liver of Pregnancy Complicated With Disseminated Intravascular Coagulation. *AACE Clinical Case Reports*, 1(4), e250-e254.
4. Karaca, Z., Tanriverdi, F., Unluhizarci, K., & Kelestimur, F. (2010). Pregnancy and pituitary disorders. *European Journal of Endocrinology*, 162(3), 453-475.
5. Goswami, R., Kochupillai, N., Crock, P. A., Jaleel, A., & Gupta, N. (2002). Pituitary autoimmunity in patients with Sheehan's syndrome. *The Journal of Clinical Endocrinology & Metabolism*, 87(9), 4137-4141.
6. Diri, H., Karaca, Z., & Tanriverdi, F. (2016). Syndrome de Sheehan: nouvelles perspectives sur une ancienne maladie. *Endocrine*, 51, 22-31.
7. Goel, A., Ch, L. C. N., Ch, E., Balasubramanian, K., & Elias, E. (2018). Acute Fatty Liver of Pregnancy Better Understanding of Pathogenesis and Earlier Clinical Recognition Results in Improved Maternal Outcomes. *EMJ Hepatol*, 6(1), 72-79.
8. Ko, H., & Yoshida, E. M. (2006). Acute fatty liver of pregnancy. *Can J Gastroenterol*, 20(1), 25-30.