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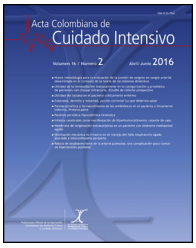

CASE REPORT

Plasmapheresis for acute liver failure in acute fatty liver of pregnancy

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KEYWORDS
Acute fatty liver of pregnancy;
Plasmapheresis;
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HELLP syndrome

Abstract Acute fatty liver of pregnancy (AFLP) is an obstetric emergency that complicates gestation in the last trimester or immediate post-partum period. It is associated with genetic defects in the long chain 3-hydroxyacyl CoA dehydrogenase (LCHAD) enzyme, which is necessary for mitochondrial fatty acids beta-oxidation, resulting in hepatic micro-vesicular steatosis and acute liver failure. Liver biopsy is the reference standard for its diagnosis, however the Swansea criteria are also used.

A case is presented of a 16-year-old woman during week 37 gestation, with biochemical evidence of acute liver failure progressing to multiple organ dysfunction. Pregnancy interruption and treatment of acute liver failure with plasmapheresis lead to the resolution of the organ dysfunction.

PALABRAS CLAVE
Hígado graso agudo del embarazo;
Plasmaféresis;
Criterios de swansea;
Síndrome HELLP

Plasmaféresis para la insuficiencia hepática aguda en el hígado graso agudo del embarazo

Resumen El hígado graso agudo del embarazo (HGAE) es una emergencia obstétrica que complica la gestación en el último trimestre o en el posparto inmediato, se asocia con defectos genéticos en la enzima 3-hidroxiacil-CoA deshidrogenasa (LCHAD) de cadena larga, necesaria para la beta-oxidación de ácidos grasos mitocondriales lo que resulta en la esteatosis micro vesicular e insuficiencia hepática aguda. La biopsia del hígado es el estándar de oro para el diagnóstico, sin embargo los criterios de Swansea también se utilizan.

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Este reporte de caso es de una mujer de 16 años de edad quien durante la semana 37 de gestación, cursó con pruebas bioquímicas de insuficiencia hepática aguda avanzando a disfunción orgánica múltiple. La interrupción del embarazo y el tratamiento de la insuficiencia hepática aguda con plasmaféresis condujeron a la resolución de la disfunción orgánica.

Introduction

Acute fatty liver of pregnancy (AFLP) is a rare complication of pregnancy with an incidence of about 5 cases per 100,000 pregnancies,[1] being an obstetric emergency with a mortality up to 90%, however nowadays with medical treatment including plasmapheresis reduces death to less than 10%.[2] AFLP is a form of severe liver disease with development of acute liver failure, jaundice, intravascular coagulation, encephalopathy, renal injury and even pancreatitis, leading to multiple organ dysfunction.

The gold standard for diagnosis is liver biopsy, which demonstrate the microvesicular steatosis, however clinical and biochemical Swansea criteria are currently used, having a negative predictive value of 100%.[3]

Treatment of acute liver failure include pregnancy interruption, blood transfusion support and more recently plasmapheresis, which in different studies is the therapeutic option with the highest success rate.[4]

Clinical case

16-Years-old female patient, without chronic diseases diagnosis. Among the gyneco-obstetric history, is the patient's first gestation denying previous pregnancies or abortions; she referred to seek monthly prenatal care without any complications.

The current condition started on June 3 of 2015 with 37 weeks of gestation, where epistaxis, gingival bleeding and asthenia characterized the clinical picture. The presence of jaundice caught the attention of the family. The first blood tests showed hyperbilirubinemia of 14.1 mg/dL and direct bilirubin of 13.2 mg/dL, being the reason why this patient was sent to the gyneco-obstetric Hospital, where was received with a biochemical data of acute liver failure and acute fetal distress, so it was decided to interrupt the pregnancy. The product was died.

The patient was transferred to intensive care 36 h after admission for further deterioration of biochemical parameters, multiple organ dysfunction with a SOFA of 18 pts, hepatic, renal and hematological failure, also showing disseminated intravascular coagulation with gastrointestinal and pulmonary bleeding, requiring invasive mechanical ventilation. Hem derivatives and factor VII were transfused, stopping the bleeding; however, the patient developed lung injury with refractory hypoxemia, needing ventilation in a prone position 12 h for 2 consecutive days (Fig. 1).

For kidney injury, slow renal replacement therapy was continuously provided for 72 h, observing renal biochemistry improvement. However, with the biochemical data of acute liver persistence, the beginning of plasmapheresis therapy was performed seven days after pregnancy interruption. After five sessions of plasmapheresis with albumin, liver failure biochemical data improved.

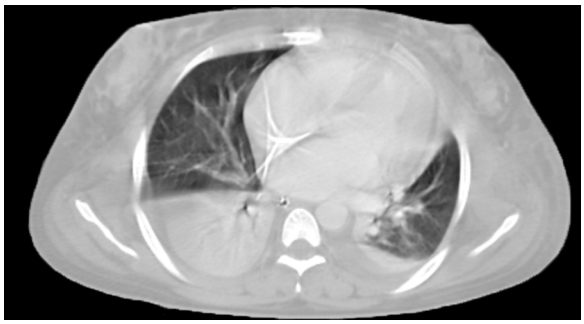

Figure 1 Thorax axial section tomography, pulmonary window. Areas of alveolar collapse and air bronchograms are observed.

For the diagnostic approach infectious causes for acute liver failure were discarded, determining them by proofs against hepatitis A, B, C and E antibodies, serological HIV, TORCH, as well as dengue virus. Among the non-infectious causes antibodies to SLE, primary biliary cirrhosis were requested, hepatic vein thrombosis was discarded, Swansea criteria were applied, concluding with a final diagnosis of pregnancy acute fatty liver (Table 1).

After 20 days of intensive care, the patient was discharged for clinical improvement with a multiple organ dysfunction diagnosis.

Discussion

AFLP is an acute mitochondrial liver disease, characterized by the presence of micro and macro vesicular steatosis on a histological level and by acute liver failure at a biochemical level; presenting hyperbilirubinemia, elevated transaminases, hypoglycemia, hyperammonemia and hyperlactatemia. Histological changes are the same as observed in Reye's syndrome and its evolution can reach hepatic necrosis or progress to liver failure, requiring a transplant.[5–7]

The main differential diagnosis of AFLP is preeclampsia and HELLP syndrome, the last one is a multisystem syndrome characterized by hypertension after 20 weeks of gestation with systolic >140 mmHg and diastolic blood pressure of 90 mmHg, associated with proteinuria of 300 mg/24 h or any other organ dysfunction as acute kidney injury, liver injury, coagulopathy or neurological disorders; the etiology resides in alterations during placentation, leading to hypo perfusion and endothelial dysfunction, where the trophoblast fails not invading the lining of the uterus, resulting in arterial placental perfusion defects getting worse as