cium channel antagonists;[8] and lymphatic vessels malformations.[9]

During the training period, chyloperitoneum was obtained that attenuated with lavage.

The suspicion of chyloperitoneum arises in the presence of milk-like fluid and the diagnosis is confirmed when chylomicrons are detected, ortriglycerides levels in the peritoneal fluid are higher than those in plasma, and protein value in the peritoneal fluid is higher than half of the protein value in plasma, like in the reported case. Differential diagnosis should be made with conditions accompanied with turbid fluid, normal cellularity and negative cultures.

A expectant attitude was taken. Regular oil was replaced by medium-chain triglycerides (MCT)-containing oil.

Treatment is usually conservative. A protein and carbohydrate rich diet is recommended with low fat content. MTC oils are preferred. The ligation of the thoracic duct can prevent nutritional complications.

Currently the patient presents intermittent and self-limited chyloperitoneum (every 10 days approximately). Nutritional parameters are preserved with daily intake of 2 liters of Nutrineal. The patient is wheel chair-bound due to FAP-associated disability.

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Pospartum hemolytic uremic syndrome: a rare entity and a treatment challenge

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Summary

Hemolytic uremic syndrome (HUS) is a rare entity that in 7% of cases has been related to oral contraceptives, pregnancy and puerperium, In this clinical setting prognosis is worse and renal replacement therapy is usually needed. Different authors agree that plamapheresis is the treatment of choice, and has improved patient survival to 80-90%. We describe a case of a young woman that 10 days postpartum developed thrombocytopenia, microangiopathic hemolytic anemia and acute renal failure with nephrotic range proteinuria. With the suspiction of HUS she was started on plasmapheresis initially stopped due to an anaphylactic reaction to plasma and finally due to hyperhidratation with acute pulmonary edema needing mechanical ventilation. Renal biopsy confirmed the diagnosis. Clinical course was complicated with refractory hypertension and infectious complications In conclusion postpartum HUS is a rare clinical entity , that forces a differential diagnosis with hypertensive complications of pregnancy. It is associated to multiple complications difficult to handle during follow-up. Plasmapheresis treatment adds complexity to clinical care but is the only treatment of proven efficacy in order to improve survival and renal prognosis.

Key words: *Hemolytic uremic syndrome. Plasmapheresis. Pregnancy. Puerperium.*

Resumen

El síndrome hemolítico urémico (SHU) es una entidad de escasa incidencia, donde un 7% de casos se asocia a la toma de anticonceptivos orales, al embarazo y al puerperio, siendo tales casos de peor pronóstico, pues frecuentemente necesitan tratamiento renal sustitutivo1. Distintos autores coinciden en que la plasmaféresis es la terapia de elección, que ha mejorado la supervivencia a 80-90%2,3. Describimos el caso de una joven que en el décimo día del puerperio presenta plaquetopenia, anemia hemolítica microangiopática (AHM), e insuficiencia renal con proteinuria nefrótica. Con la orientación de SHU se pauta plasmaféresis, que se suspende inicialmente por alergia al plasma infundido, y definitivamente por hiperhidratación con edema agudo de pulmón (EAP) que precisa ventilación mecánica. La biopsia renal confirma el diagnóstico de presunción. La evolución resulta tórpida, marcada por la hipertensión arterial (HTA) refractaria y complicaciones infecciosas. En conclusión, el SHU post-parto es una patología poco frecuente, que asocia muchas complicaciones de difícil manejo a lo largo de su evolución y que obliga al diagnóstico diferencial con los estados hipertensivos del embarazo4. A su vez, el tratamiento con plasmaféresis añade complejidad al cuadro, pero es el único procedimiento que ha demostrado mejorar la supervivencia y el pronóstico renal.

Palabras clave: *Sindrome hemolítico urémico. Plasmaféresis. Embarazo.*

To the editor: The hemolytic uremic syndrome (HUS) is an acute and potentially fatal form of thrombotic microangiopathy, with an incidence of 17.2 new cases/per million population/year. Seven percent of the cases are associated to oral contraceptive treatment, pregnancy and puerperium. Puerperium-related cases have the worst prognosis, as renal replacement treatment is often required.[1] The plasmapheresis is elective, as it is the only therapy, that has shown to improve the survival by 80%-90%[2,3] and the prognosis of the renal involvement.

We present a 20 year-old Caucasian woman in her first pregnancy that underwent a cesarean at 41st week because the delivery did not progress. The baby was a healthy male. On the 10th day after the delivery she was evaluated because of lumbar and hypogastric pain

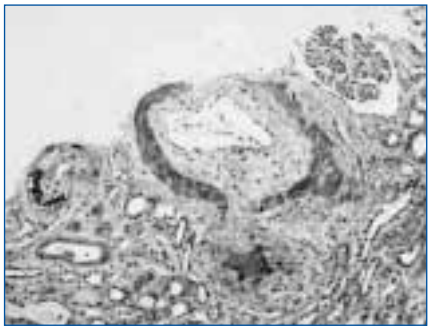

Figure 1.

and fever of 38.5 ºC. Blood parameters were: urea 224.6 mg/dL, creatinine 6.46 mg/dL, albumin 2.9 g/dL, hemoglobin 5.9 g/dL, 16% of schistocytes in peripheral blood, haptoglobin undetectable, LDH 4.162 U/L, and platelet count 104 x 10^9/L. The urinalysis showed proteinuria 20 g/24-hours with normal sediment. D-dimers determination was 16.381 mcg/L and for this reason an MRI without contrast was performed that ruled out renal vein thrombosis. The gynecologic exam showed placental remnants in the uterus, she was diagnosed with endometriosis and a uterine scraping was performed.

Sixty-two to seventy-four percent of pregnancy-related HUS cases occur at the end of the pregnancy. Differential diagnosis should be made with hypertensive conditions of the pregnancy, which produce symptoms that do not last more than 3 days after the delivery. Persistent or worsening symptoms beyond the third day of the puerperium decrease the probability of spontaneous recovery and strength the indication of plasmapheresis.[5] The reported patient was on the tenth day after the delivery, and daily plasmapheresis was initiated, which had to be discontinued, first because of allergy to infused plasma and definitively because of acute pulmonary edema that prompted mechanical ventilation.

The patient had severe hypertension, which required five drugs to be controlled. The reason could be found in the pathological study of the renal biopsy that revealed lesions typical of thrombotic microangiopathy, with severe vascular involvement, including intimal proliferation and hyperplasia, presence of glomeruloid bodies and several glomerular changes. It is known that patients with HUS and arterial involvement (with no prodromes such as bloody diarrhea), as in the present case, have a worse prognosis than those who only have glomerular involvement (and prodromal bloody diarrhea).[6]

Despite appropriate treatment renal function was not recovered and the patient required chronic hemodialysis. The risk of recurrence in further pregnancies is high, and chronic renal failure is the most important consequence, which happens in 25% of the cases.

The pathophysiological hypothesis is a deficiency of the ADAMTS13 proteinase, which degrades the serum polymers of von Willebrand factor, due to the high levels of estrogens during the pregnancy, particularly at weeks 36-40.[8,9] Anyway we cannot rule out that the endometriosis could be an infectious cause of HUS in this patient.

This case illustrates the difficulty to manage HUS in the puerperium, especially regarding the differential diagnosis and management.

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