The main approach to this condition should be the prevention, through monitoring of calcium levels and appropriate frequency of dialysis sessions. In experimental models it has been shown that early use of biphosphonates decreases the incidence.[9] Parathyroidectomy is only beneficial in case of very PTH high levels.[10]

In the literature review we have found 35 cases of calciphylaxis with penis involvement.[2,11,12] In 35% of them the treatment was conservative, in 53% surgery was underwent only if complications developed and in 12% surgery was early performed. The mortality rate was 58%, 61% and 25%, respectively. Sixty-eight percent of the cases evolved to moist gangrene (2,11). If the penis is involved the mortality reaches 69%.[2,12]

A high mortality of 50% at 6 months has been reported when penis necrosis develops in patients with DM and end-stage CRF.[13] DM, high blood pressure, end-stage CRF and dyslipidemia accelerate the atherosclerotic angiopathy, and they are the main risk factors for this entity.

Penis calciphylaxis is an infrequent systemic presentation of end-stage CRF. Early diagnosis and appropriate management can be determinant for the evolution and the prognosis in these patients.

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Chyloperitoneum and amyloidosis

Nefrología 2008; 28 **(1)** 119-120

Summary

Chyloperitoneum is very infrequent in peritoneal dialysis. It has been described in patients diagnosed of systemic amyloidosis. We present the case of a patient in program of peritoneal dialysis, with familial amyloidosis type Andrade disease that presented intermittent and recidivante chyloperitoneum.

Key words: *Chyloperitoneum. Peritoneal dialysis. Amyloidosis.*

Resumen

El quiloperitoneo es una entidad rara en diálisis peritoneal. Se ha descrito en pacientes diagnosticados de amiloidosis sistémica. Presentamos el caso de una paciente en programa de diálisis peritoneal, con amiloidosis familiar tipo enfermedad de Andrade que cursó con quiloperitoneo intermitente y recidivante.

Palabras clave: *Quiloperitoneo. Diálisis peritoneal. Amiloidosis.*

To the editor: Chyloperitoneum is a rare condition in patients on peritoneal dialysis.[1] It has been reported in patients diagnosed with systemic amyloidosis. We present a patient on peritoneal dialysis, with familial amyloidosis Andrade type with intermittent and recurrent chyloperitoneum.

The patient was a 69 year-old woman who began peritoneal dialysis in April of 2006. She had renal amyloidosis in the setting of familial amyloid polyneuropathy (FAP), which had manifested with nephrotic syndrome.

The clinical picture onset at 64 year-old with hyperestesia of the lower limbs. The electromyography was compatible with mixed polyneuropathy. She had the transthyretin transthyretin (TTR) mutation (Val-30-Met). Rectal biopsy study showed amyloid deposition, and the diagnosis of FAP was confirmed. At presentation, renal function was normal and the proteinuria was negative. She required a pacemaker because of cardiac amyloid infiltration.

Mallorca is the fifth world endemic focus of FAP or Andrade's disease,[2] a hereditary systemic amyloidosis, with mixed polyneuropathy being the main manifestation. The inheritance is autosomal dominant. The disease is due to a transthyretin mutation, which is synthesized in the liver and forms systemic depositions, which cause the clinical picture.

The diagnosis is made by means of rectal, fat or sural nerve biopsy, that discloses the TTR variant or the DNA mutation.

The only efficacious treatment is liver transplantation.

In January of 2004 proteinuria appeared, which reached nephrotic range. Since April of 2005, the renal function deteriorated progressively, and in February of 2006 a catheter for peritoneal dialysis was placed.

During peritoneal catheter implantation peritoneal lavage was performed and a white fluid was obtained.

Chyloperitoneum is the appearance of a turbid milk-like fluid due to the presence of chylomicrons. Among the causes that can be implicated are: micro-traumas, sometimes during catheter insertion;[3] malignant diseases, mainly lymphomas;[4] hepatic cirrhosis, acute pancreatitis;[5] systemic amyloidosis[6] as in the case reported; lymphatic obstruction due to fibrotic adherences of tuberculosis;[7] dihydropiridinic cal-

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

Nefrología 2008; 28 **(1)** 120-121

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