



Figure 1.

A 67 year-old male came to the Emergency Room of a hospital in Cali, Colombia, because of hiccups for 2 days. He had a history of type 2 diabetes mellitus with secondary renal failure and was on hemodialysis three times per week for 19 months. Twelve hours before his first visit, he had taken star fruit juice in a quantity equivalent to 4 pieces of fruit. In the Emergency Room no apparent cause for the hiccups was found and the patient was referred to our center for the scheduled hemodialysis, after which the hiccups disappeared. He went home and drank a similar quantity of juice. Almost immediately the hiccups recurred followed by vomiting and the patient was admitted to the hospital. During the two following days the patient presented progressive neurological deterioration leading to stupor.

The patient was again referred to our center. On admission he was stuporous, with no neurological focalization and irregular respiratory pattern. The physical exam was otherwise unremarkable. Laboratory investigations showed mild anemia, glycemia 315 mg/dL (17.5 mmol/L), and creatinine 3.93 mg/dL (347.4 mmol/L). Cranial images were normal. The association between the clinical picture and the star fruit intake was evident. The patient remained for the following six days at the intensive care unit, and daily hemodialysis with 4 hours sessions was performed. The symptoms disappeared progressively with no consequences, except for a complete amnesia of the facts that happened around the intoxication episode. Since then he has never again taken the fruit. At the Emergency Room, a presentation with hiccups should arise the suspicion of this potentially fatal in-

toxication in patients with chronic renal disease.

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### Penis necrosis as an unusual manifestation of calciphylaxis in uremia

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**To the editor:** Penile necrosis is a rare entity and only a few cases have been described in patients on dialysis.<sup>1,2</sup> In these patients it can be associated to diabetes mellitus (DM), cholesterol emboli and seldom to calciphylaxis.<sup>1,3,4</sup> The diagnosis relies on clinical picture, history, physical exam and on other investigations like skin biopsy.<sup>1</sup> We present a case of calciphylaxis in an infrequent location in a patient on hemodialysis.

A 43 year-old male was on periodic hemodialysis because of chronic glomerulonephritis. In the next three years the Ca/P product was persistently higher than 70 mg<sup>2</sup>/dL<sup>2</sup>, he had hyperphosphatemia and severe hyperparathyroidism (iPTH 1200 pg/mL). No control could be achieved either with calcitriol or with subtotal parathyroidectomy, which was performed in January of 2005. Since the intervention the PTH level was 200 pg/mL, and calcium and phosphorus levels were 8.5 mg/dL and 4 mg/dL, respectively. In September of 2005 he was

admitted because of general deterioration and progressive appearance of petechiae on the penis, which evolved to necrosis (fig. 1).

Laboratory findings were the following: 9,100 leucocytes/mm<sup>3</sup>, Hb 9.8 g/dL, proteins 4.4 g/dL, P 4.32 mg/dL, and PTH 248 pg/mL. Coagulation parameters were normal and HIV serology was negative. A plain X-ray film of the abdomen showed calcifications within the iliofemoral vessels. Doppler ultrasound of the penis and the iliac vessels revealed a complete lack of flow in the penis and minimal flow in the iliac and femoral arteries, as well as calcifications within the penis vessels.

A decision to perform a partial penectomy was made. The pathological study disclosed hyperplasia of the intimal layer, calcifications in the media with necrotic areas and bleeding within the penis. The postsurgical evolution was acceptable.

Calciphylaxis is a disorder of unknown etiopathogenesis. It is associated to hypercalcemia and/or hyperphosphatemia due to secondary hyperparathyroidism or to intake of calcium preparations and calcitriol.<sup>5</sup> It appears in 1%-4% of the patients on hemodialysis. It was also described in patients on peritoneal dialysis, and very rarely in patients with renal transplantation or in those with end stage CRF, stages III and IV.<sup>6-8</sup>

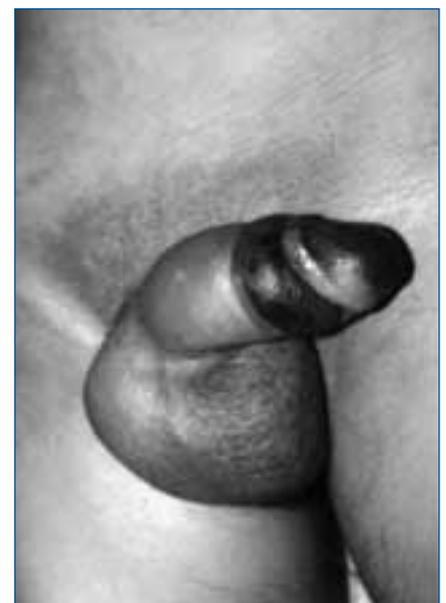


Figure 1. Lesion of the glans penis with a necrotic appearance.

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 bisphosphonates decreases the incidence.<sup>9</sup> Parathyroidectomy is only beneficial in case of very PTH high levels.<sup>10</sup>

In the literature review we have found 35 cases of calciphylaxis with penis involvement.<sup>2,11,12</sup> 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%.<sup>2,12</sup>

A high mortality of 50% at 6 months has been reported when penis necrosis develops in patients with DM and end-stage CRF.<sup>13</sup> 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

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### 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 quilooperitoneo 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 quilooperitoneo intermitente y recidivante.**

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

**To the editor:** Chyloperitoneum is a rare condition in patients on peritoneal dialysis.<sup>1</sup> It has been reported in pa-

tients 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 hyperesthesia 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,<sup>2</sup> 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;<sup>3</sup> malignant diseases, mainly lymphomas;<sup>4</sup> hepatic cirrhosis, acute pancreatitis;<sup>5</sup> systemic amyloidosis<sup>6</sup> as in the case reported; lymphatic obstruction due to fibrotic adhesions of tuberculosis;<sup>7</sup> dihydropyridinic cal-