Dear Sir,

Chronic pyelonephritis with staghorn calculus formation is a frequent cause of chronic renal failure [1]. On some occasions, these patients are included in a dialysis program without a satisfactory urological treatment which can be the cause of occult septic foci. Although the relationship between chronic infection and amyloidosis type AA is well known [2], the association between chronic pyelonephritis and amyloidosis has scarcely been reported in patients without spinal cord injury [3, 4].

We present the cases of 3 female patients diagnosed as having chronic renal failure secondary to chronic interstitial nephritis with staghorn calculi, who developed a systemic amyloidosis soon after they had begun a chronic hemodialysis treatment.

Case 1

A 66-year-old female with a previous history of frequent urinary tract infections and lithiasis was remitted to our Service to study a renal failure. She had no history of any other inflammatory or infectious diseases. In the study, an end-stage renal disease was confirmed by a morphological study that showed an atrophic post-pyelonephritic right kidney and a left kidney with postobstructive cortical atrophy and two large staghorn calculi, one in the pelvis and the other in the proximal ureter. There were no hepatic abnormalities, and an urinary culture yeilded Proteus mirabilis. The 24-hour urinary protein excretion was 1.2 g.

The patient was included in hemodialysis with an uneventful course, save recurrent symptomatic urinary tract infections that responded well to antibiotic therapy. Ten months later, the analytical test began to show a moderate elevation of γ-glutamyltranspeptidase and alkaline phosphatase with mild increments in transami-nases, having ruled out infections of hepatitis A and B viruses. These alterations increased in the following 2 months. A hepatic biopsy was performed, showing a diffuse hepatic infiltration sensible to amyloid permanganate. The patient died suddenly 2 months later due to a presumably ventricular arrhythmia.

Case 2
A 50-year-old female with antecedents of two surgical procedures for staghorn calculi 10 and 8 years ago was remitted from the Urology Service because of renal insufficiency. The study showed a creatinine level of 397 µmol/l and creatinine clearance of 0.150 ml/s. By sonography the right kidney was atrophic, and the left showed a posthydronephrotic atrophy with a large staghorn calculus that occupied the pelvis and calyces. A urinary culture yielded a Proteus, and the urine presented a mild proteinuria (0.5 g/24 h). Three months later the patient was included in a hemodialysis program, presenting from the beginning vomiting and diarrhea. A colonoscopic examination showed a friable and bloody colonic mucous membrane. The histological examination revealed the presence of Congo-red-positive material in the basal membrane and vessels permanganate-sensible. The patient suffered a stormy clinical course with starvation diarrhea and died 40 days later due to sepsis.

Case 3

A 49-year-old female, diagnosed as having bilateral staghorn calculi and advanced renal failure, was remitted to our Service. In the study, we confirmed the renal insufficiency (creatinine 468 µmol/l and creatinine clearance 0.133 ml/s). In the urine a mild proteinuria (0.7 mg/24 h) and a positive culture of Escherichia coli were observed. She had no history of other infectious or inflammatory disorders. By sonography, a large obstructive staghorn calculus was observed in the left kidney with atrophy of the cortex. The right kidney was atrophic with a nonobstructive staghorn calculus. She was included in a hemodialysis program 1 month later. The patient was warned of the necessity of bilateral nephrectomy to access to a transplant program, but she refused it. Sixteen months later, the patient began with watery diarrhea and weight loss. The diarrhea became mucous and bloody. A colonoscopy was performed, showing an ulcerative aspect of the colonic mucous membrane similar to Chronic Pyelonephritis and Systemic Amyloidosis.

an ulcerative colitis. The biopsy showed a diffuse amyloid infiltration type AA (Congo-red- and thioflavin-T-positive staining, permanganate-sensible), but without changes suggesting ulcerative colitis. An anti-β2-microglobulin monoclonal antibody staining of the sample was negative. Because of septic manifestation during her hospitalization and the sonographic suspicion of left pyonephrosis, she was nephrectomized. The kidney showed a collection of 200 ml of purulent material from which a culture yielded E.coli. The parenchyma was inflammatory and atrophic, but there were no findings of xanthogranulomatous pyelonephritis. Glomeruli and vessels showed a positive staining of Congo red and thioflavin T. The patient died 15 days later because of septic shock.

The 3 patients described showed very similar characteristics in their clinical picture: presumably long-term obstructed and septic kidney and an end-stage renal failure. Although no patient presented suspected symptoms of amyloidosis prior to dialysis treatment, actually we cannot know to what extent the amyloidosis played a role in the development of renal failure. Two cases of amyloidosis AA associated with xanthogranulomatous pyelonephritis have previously been reported [3,4]. Xanthogranulomatous pyelonephritis was pathologically ruled out in 1 of our patients, and in the other 2 there were no morphological data to suspect it. For that, we think that the link between xanthogranulomatous pyelonephritis and amyloidosis is the common pathogenic ways, that is obstruction and infection of the kidney. Amyloidosis secondary to deposition of β2-microglobulin fibrils has increasingly been reported in patients on long-term hemodialysis [5]. This amyloidosis is undistinguishable from the AA type by the permanganate...
reaction [6]. In 1 of our patients, β2-microglobulin deposition was discarded by a specific
method (anti-β2-microglobulin staining) and, although in the other 2 it was not done, the
development of a β2-microglobulin amyloidosis in such short-term hemodialysis seems highly
improbable.
In conclusion, the presence or development of amyloidosis type AA in patients with chronic
interstitial nephritis and staghorn calculi without a successful urological does not seem
infrequent. Owing to the poor prognosis showed in these 3 patients and the hypothetical
possibility to stop the progression of amyloid deposit disease, we suggest that in these patients a
more aggressive intervention should be taken even in those not included in transplant programs.
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