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Emhysematous pyelonephritis and polycystic hepatokidney radical nephrectomy

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Dear Editor,

Adult polycystic kidney disease (PCKD) is an inherited disorder characterised by the presence of kidney cysts, which affects approximately one in every 500 to 1,000 people. The most common manifestations are lumbar pain, haematuria and recurrent urinary infections.[1,2] Emphysematous pyelonephritis is a severe infection characterised by the presence of gas in the kidney parenchyma, collecting system or perinephric tissue, and occurs primarily in diabetic patients.[3]

We report the case of a 76 year-old woman with hypertension, diabetes, chronic kidney disease secondary to polycystic hepatokidney disease in a peritoneal dialysis programme since March 2008, when she was admitted for fever, dysuria and haematuria. Laboratory tests showed elevated inflammatory parameters and the presence of *E. coli* from urine and blood culture tests. A CT scan showed a cyst on the upper right kidney complicated with air in the calices and bladder without prior manipulation of the urinary tract (Figure 1). Broad-spectrum antibiotic treatment with meropenem and gentamicin, according to the antibiogram sensitivity, was started and the patient was temporarily transferred to haemodialysis to treat intercurrent septic profile better. Despite this treatment, the patient continued to be febrile and showed a deterioration in her general condition. A new tomographic control was therefore done and a deterioration of the right kidney cyst was seen. Given the poor outcome of the septic profile with a lack of response to treatment, a right radical nephrectomy was performed, which revealed multiple kidney cysts filled with pus (Fig. 2). Further evolution was slow towards the improvement of her general condition, with a practical resolution of fever and progressive normalisation of the inflammatory parameters. Finally, the patient was able to be released after a long period of convalescence in a clinical and haemodynamically stable state, continuing kidney replacement therapy with regular haemodialysis.

In PCKD, recurrent urinary tract infections can cause septic conditions which are difficult to control due to secondary infection of the cysts. The prevalence of cystic infection is high (30-50%), and is more common in women with a history of urinary tract manipulation, nephrolithiasis and/or vesicoureteral reflux. The quinolones, trimethoprim/sulfamethoxazole and chloramphenicol reach good therapeutic concentrations within the cyst. A good response to antibiotic treatment is observed in most cases, and it is rarely necessary to take aggressive interventionist measures such as percutaneous drainage, and much less frequently, nephrectomy.

Emphysematous pyelonephritis is a severe kidney infection, usually caused by gas-producing coliform bacteria, with *E. coli* the most common organism, usually affecting patients with diabetes. It is characterised by its aggressive progression and poor response to treatment. It is associated with a high mortality, so nephrectomy may be advisable in selected cases.

A better understanding of the disease, the vast antimicrobial arsenal available today, with greater synergy and antibiotic potency, the emergence of minimally invasive interventional radiologic techniques and the fact that nephrectomy has a significant rate of morbidity and mortality (12 and 5%, respectively) in high-risk surgical patients, are among the factors to be considered for nephrectomy in patients with terminal kidney disease.[3-5] At present, radical or partial nephrectomy should be considered for patients with a poor response to antibiotic treatment or percutaneous drainage due to its potential complications.

To summarise, the above is an example of a patient with emphysematous pyelonephritis and hepatokidney polycystosis with, firstly, a slow evolution of an unusual and severe kidney infection and, secondly, the need for radical nephrectomy as definitive treatment.

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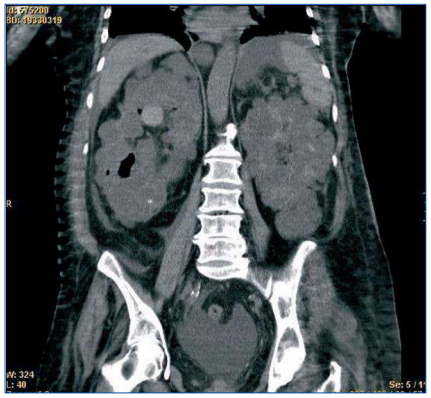

Figure 1. Abdominal CT scan: coronal with complicated cyst in upper third right kidney and air in the urinary tract.

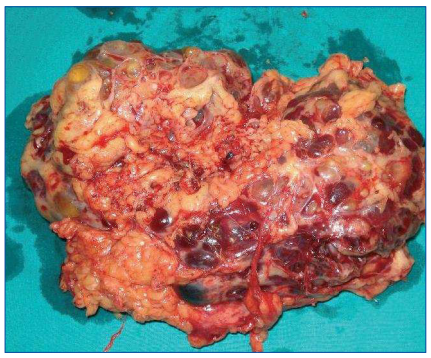

Figure 2. Surgical specimen: polycystic right kidney 20x14cm, 4kg, with multiple complicated cysts with pus.

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A case of IgD lambda multiple myeloma with acute kidney injury

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Dear Editor,

Monoclonal gammopathies include multiple clinical entities characterised by clonal proliferation of plasma cells that produce a homogeneous monoclonal paraprotein.[1] To identify this protein serum immunoelectrophoresis or immunofixation need to be used.[1] These monoclonal gammopathies can be classified as malignant, uncertain, idiopathic or transitory (the latter can occur in patients undergoing kidney transplantation due to immunosuppressive therapy, especially in older age).[1] Multiple myeloma or Kahler's disease is the prototype of malignant monoclonal gammopathy and is the most frequent neoplasm in plasma cells.[1] It occurs mainly in elderly males (60-65 years) and its clinical features include bone, kidney, neurological, and, of course, haematological disease. Kidney function is impaired in 50% of patients at the time of diagnosis.[2] It may be seen with tubular and/or vascular glomerular injury.[2] Kidney involvement is the single most influential factor in poor prognosis and in 50% of cases is triggered by hypercalcaemia.[1] The second mechanism of kidney injury is due to excessive production of immunoglobulins and/or light chains (Bence-Jones proteinuria) which are filtered by the glomeruli and saturate the kidney's ability to absorb by the proximal tubule and catabolise in the lysosomes.[2]

We report the case of a man aged 84 who attended emergency due to poor health, anorexia and weight loss during a month. His background revealed a history of hypertension and coronary artery disease. The physical examination showed a tendency to hypotension with discrete signs of dehydration without other relevant findings. The blood test gave: Hb 8.9g/dL, platelets 67,000L, leukocytes 7,700 per µL, creatinine 12mg/dL, urea 250mg/dL, K 6.7mEq/L, HCO_3 9mEq/L. Abdominal ultrasound displayed both kidneys of 12cm with good corticomedullary differentiation and without signs of obstructive uropathy. On one occasion, a proteinogram in blood and urine was requested in the ward, which detected an IgD lambda monoclonal peak in blood of 0.89g/dL and urine of 1.68g/24 h. 60-90% infiltration of plasma cells was detected in the bone marrow study. Chemotherapy with bortezomib, melphalan and steroids is prescribed after diagnosis of IgD myeloma with cast nephropathy (or myeloma kidney). Inclusion in a haemodialysis programme was requested urgently. After 6 months, there was no objective improvement of kidney function.

This case is interesting not so much for pathology, but for one of its subclasses. IgD monoclonal gammopathy represents 1% of all myelomas, compared with IgG and IgA myelomas, which constitute 53 and 28% respectively of the total.[1] The IgD myeloma takes a characteristically more aggressive course, and is associated with increased kidney disease rate (virtually 100% of the cases have Bence-Jones proteinuria).[3,5] Myeloma clinical suspicion must always be present in the differential diagnosis of any cases of acute kidney injury, especially in elderly patients with a history of constitutional syndrome.

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