

Idiopathic fibrosis as the cause of obstructive uropathy

Nefrologia 2013;33(3):439-40

doi:10.3265/Nefrologia.pre2012.Sep.11705

To the Editor:

Retroperitoneal fibrosis represents a diagnostic challenge due to the low frequency of presentation and the lack of specific symptoms. It is characterised by the presence of fibro-inflammatory tissue that surrounds the abdominal aorta and the iliac arteries¹ extending to the retroperitoneal space and it covers neighbouring structures, such as ureters, causing obstructive uropathy (OU).

Although cases associated with multiple conditions (drugs, neoplasias, radiation and infection, amongst others) have been described, the most common cause is idiopathic.²

Idiopathic retroperitoneal fibrosis is more common in men aged 50-60 years old and, in most patients, it presents as non-specific lower back or abdominal pain.³

When the retroperitoneal tissue covers one or both ureters, potentially reversible obstructive renal failure occurs.^{1,2,4}

We report the case of a patient with idiopathic retroperitoneal fibrosis that progressed to marked OU and an ultrasound showed slight unilateral dilation of the excretory system.

Our patient was a 54-year-old male, with no relevant personal or family history, referred by the General Practitioner due to incidental findings in the blood test of creatinine 2.74mg/dl. The patient had a history of three weeks of nausea, non-specific abdominal pain and slight testicular pain, with no

other accompanying symptoms. Upon physical examination, he was normotensive and afebrile, with diuresis maintained, without macroscopic abnormalities, at around 2000ml/day.

In the blood test obtained from the Emergency department, the following parameters are notable: haemoglobin 15.3g/dl, leukocytes 6700/ μ l, platelets 305,000/ μ l, sodium 133mEq/l, potassium 3.6mEq/l, hepatic-pancreatic profile with no abnormalities, lactate hydrogenase 270U/l, plasma creatinine 3mg/dl, urea 116mg/dl. Urine biochemistry displayed values for sodium of 96mEq/l and potassium of 28mEq/l.

The renal ultrasound (Figure 1), showed normal-sized kidneys with cortical thickness maintained and we noted that the right side was larger and thicker. It showed slight dilatation of the right renal calyces (Figure 1) and no cause of dilation was seen with a bilateral positive ureteral jet on colour Doppler ultrasound.

Given the poor test performance with progressive deterioration of renal function, a computerised tomography (CT) scan of the abdomen was carried out (Figure 2). It revealed a large retroperitoneal mass that covered the right ureter, with the left ureter free, but on the limit of the edge of the latter, resulting in bilateral OU, although it was more pronounced on the right side. The left side only showed incipient signs of obstruction.

After the diagnosis, he was treated empirically with high doses of prednisone and it was decided to use bilateral double-J catheters; following this treatment immediate improvement was noted. Five days later, the patient was asymptomatic and 30 days after starting steroid treatment, plasma creatinine was 1.02mg/dl. The double-J catheters

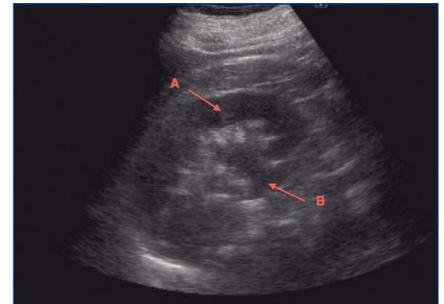


Figure 1. Right kidney ultrasound. The kidney is a normal size, with cortical thickness maintained, with corticomedullary differentiation (A) and slightly dilated renal calyces (B).

were removed 6 months after the start of treatment, with stable plasma creatinine figures being maintained.

This case is of interest because, in spite of being an obstructive disease, ultrasound does not make clear the cause of obstruction, and on many occasions it fails to reveal the existence of the urinary tract obstruction itself. This is due to ureteral rigidity that conditions the disease itself, preventing dilation of the ureter covered by fibro-inflammatory mass.

In the case that we have presented,



Figure 2. Computerised tomography. A computerised tomography scan displaying the retroperitoneal mass covering the right ureter (A). The left ureter is free, but close to the limit of the retroperitoneal mass (B).

the patient displayed a significant impairment in renal function secondary to bilateral OU (right established and left incipient), with CT being required for its diagnosis.

Therefore, it is necessary to evaluate the need for other imaging techniques such as CT or MRI,⁵ in order to diagnose this entity, since treatment started at an early stage improves the clinical profile.

Conflicts of interest

The authors declare that they have no conflicts of interest related to the contents of this article.

1. Kermani TA, Crowson CS, Achenbach SJ, Luthra HS. Idiopathic retroperitoneal fibrosis: A retrospective review of clinical presentation, treatment and outcomes. *Mayo Clin Proc* 2011;86(4):297-303.
2. Vaglio A, Salvarani C, Buzio C. Retroperitoneal fibrosis. *Lancet* 2006;367(9506):241-51.
3. Corradi D, Maestri R, Palmisano A, Bosio S, Greco P, Manenti L, et al. Idiopathic retroperitoneal fibrosis: clinicopathologic features and differential diagnosis. *Kidney Int* 2007;72(6):742-53.
4. Rodríguez Jornet A, Andreu Navarro FJ, Orellana Fernández R, Ibeas López J, García García M. Idiopathic retroperitoneal fibrosis: clinicopathological characteristics. *Nefrologia* 2009;29(4):298-303.
5. Scheel PJ Jr, Feeley N. Retroperitoneal fibrosis: the clinical, laboratory and radiographic presentation. *Medicine (Baltimore)* 2009;88(4):202-7.

Alba Santos, Borja Quiroga, Nicolás Macías, Marian Goicoechea, Soledad García de Vinuesa

Sección de Nefrología.

Hospital General Universitario Gregorio Marañón. Madrid. (Spain).

Correspondence: Alba Santos

Sección de Nefrología.

Hospital General Universitario Gregorio Marañón. Madrid. (Spain).

alba.santos@salud.madrid.org

Renal autotransplantation in renovascular hypertension secondary to renal artery aneurysm

Nefrologia 2013;33(3):440-2

doi:10.3265/Nefrologia.pre2012.Nov.11825

To the Editor:

Fibromuscular dysplasia (FMD) is occasionally accompanied by renal artery aneurysms. Percutaneous treatment is the treatment of choice in simple aneurysms and surgical treatment is reserved for complicated aneurysms and other selected cases.¹

We report the case of a 36-year-old male, smoker, known as hypertensive for 20 years, who was monitored in our Hypertension Unit. During the year in which he was monitored, he had hypertensive emergency associated with headaches. Among the additional tests conducted, laboratory and eye fundus tests and renal ultrasound were normal. The echocardiography revealed a mild left ventricular hypertrophy with grade I diastolic dysfunction and the captopril test did not suggest renovascular hypertension (RVHT).

Five years later, he was admitted due to an episode of transient ischaemic attack in the context of a hypertensive emergency (210/130mmHg). At that time, the laboratory tests and renal ultrasound were normal and the eye fundus revealed a Keith-Wagen-er grade I hypertensive retinopathy. Since he was a young male with sustained high blood pressure (HBP) and frequent hypertensive emergencies, we wanted to rule out RVHT. The patient refused all forms of intervention and it was decided to continue ambulatory monitoring; optimal control of blood pressure (BP) was achieved with five drugs (irbesartan 300mg/day, hydrochlorothiazide 12.5mg/day, ramipril 5mg/12h, am-

lodipine 15mg/day, carvedilol 12.5mg/12h).

The following year, he suffered a subarachnoid haemorrhage after rupture of an aneurysm in the right superior cerebellar artery, making a full recovery after embolisation.

Subsequently, we observed a significant deterioration in renal function: creatinine (Cr) 3.7mg/dl, creatinine clearance (CrCl) 26.35 ml/min, which partially improved after discontinuation of the angiotensin converting enzyme inhibitor and the angiotensin II receptor antagonist. Given the suspicion of RVHT, a magnetic resonance angiogram was performed on the abdomen, confirming the presence of right renal artery stenosis 11mm from the ostium and an ovoid aneurysm in the final portion of the left renal artery, with a diameter greater than 1.8cm. Subsequently, renal arteriography was performed, in which we observed bilateral renal artery stenosis of probable fibromuscular dysplasia origin (Figure 1). Given these findings, bilateral percutaneous transluminal angioplasty was performed with an integral recovery of renal function: Cr 1mg/dl, CrCl 80ml/min and BP controlled with triple antihypertensive therapy (labetalol 300mg/day, amlodipine besylate 10mg/12h, torasemide 5mg/day).

Five years later, a follow-up renal ultrasound showed renal asymmetry, not previously observed (left kidney 8.5cm, right kidney 10cm), and therefore a computed tomography angiogram of the renal arteries was requested, confirming left renal artery restenosis, and the presence of an aneurysm on the right kidney of 1.5 x 1.2cm and the known aneurysm on the left side in an hourglass shape whose largest end measured 1 x 1.5 x 2.4cm. Furthermore, the ostium of 4 arterial branches was observed on the left side of the aneurysm. Given these findings, because it is an aneurysm with a complex anatomy, we ruled out the possibility of percutaneous treatment and the patient was taken to the Hospital Juan Canale-