Caso reporte

Retroperitoneal fibrosis as a cause of obstructive uropathy. Case report

Fibrosis retroperitoneal como causa de uropatía obstructiva. Reporte de caso

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Abstract

Retroperitoneal fibrosis is a rare, in most cases idiopathic, pathology, although it has been associated with medications, neoplasms and other connective tissue diseases. In terms of histopathology, inflammation and deposits of fibrotic tissue in the retroperitoneum are observed and, characteristically, this covers the ureters, provoking acute obstructive kidney damage, the most frequent manifestation of the disease. The definitive diagnosis is obtained solely via biopsy, and the basis of treatment is corticotherapy, although in severe cases, and where resistance to corticosteroids exists, other treatments have been used, such as immunomodulators. Occasionally, surgical interventions are necessary to manage complications. The case of a 50-year-old man who came to the Hospital emergency service due to abdominal pain is presented. Paraclinical studies showed azotemia, and diagnostic images showed left hydronephrosis with a component of interaortocaval and periaortic soft tissue. A retroperitoneal biopsy was conducted, and a diagnosis of idiopathic retroperitoneal fibrosis was made. Bilateral nephrostomies were put in place and treatment with corticosteroids was initiated.

Key words: Retroperitoneal fibrosis, obstructive acute kidney injury, corticosteroids, nephrostomies, Low Back Pain, hydronephrosis.

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Resumen

La fibrosis retroperitoneal es una patología rara, en la mayoría de los casos idiopática, aunque se ha asociado a medicamentos, neoplasias y otras enfermedades de tejido conectivo. Histopatológicamente se evidencia inflamación y depósito de tejido fibrótico en el retroperitoneo y se caracteriza por cubrir los uréteres provocando lesión renal aguda obstructiva siendo ésta, la manifestación más frecuente; el diagnóstico definitivo se obtiene únicamente con biopsia y la base del tratamiento es la corticoterapia, aunque en casos severos y en resistencia a los corticoides se han usado otras terapias como los inmunomuladores. En ocasiones son necesarias las intervenciones quirúrgicas para el manejo de las complicaciones. Se presenta el caso de un hombre de 50 años que ingresó al servicio de urgencias del Hospital San José por dolor abdominal, los paraclínicos demostraron elevación de los azoados y en las imágenes diagnósticas hidronefrosis izquierda con componente de tejido blando interaortocava y periaórtico, se realizó biopsia retroperitoneal y se hizo diagnóstico de fibrosis retroperitoneal idiopática, se instauraron nefrostomías bilaterales y se inició manejo con corticoide.

Palabras clave: fibrosis retroperitoneal, lesión renal aguda obstructiva, corticoides, nefrostomías, dolor de la región lumbar, hidronefrosis.

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Introduction

Idiopathic retroperitoneal fibrosis (IRF) is a rare condition, first described in 1905 by Albarra, a French urologist, and later in 1948 by Dr. John Ormond, both described cases of ureteral obstruction.[1] In 2010 it was included within the spectrum of immunoglobulin 4 (IgG 4)-related diseases, which are characterized by the accumulation of fibroinflammatory tissue in the retroperitoneal soft tissues and other abdominal organs; the tissue is most frequently found peripheral to the abdominal aorta, the iliac arteries, the urinary tract and the renal arteries. The ureters are usually trapped in this fibrous tissue, this being the cause of obstructive uropathy. It has been reported an incidence of 0.1 per 100,000 inhabitants and a prevalence of 1.4 per 100,000, the average age of diagnosis is 50 years and it occurs 2 to 3 times more in men than in women.[2]

Below, a description is made of a patient with retroperitoneal fibrosis who was referred to the San José Hospital for evidence of bilateral hydronephrosis conditioning an acute post-renal kidney injury, elevation of nitrogen compounds and oligoanuria.

Case presentation

A 50-year-old male patient with a pathological antecedent of urolithiasis for 25 years, consulted for the first time in the emergency service in 2016 due to a clinical picture of abdominal pain radiated to the lumbar region, for this reason, it was performed a simple abdominal tomography that showed bilateral renal lithiasis, left proximal ureteral lithiasis conditioning a mild left ureterohydronephrosis and additionally a component of interaortocaval and periaortic soft tissue associated with lymph node conglomerate of lymphoproliferative neoplastic nature, which is why it was decided to take an abdominal CT scan with contrast, which showed conglomerates of retroperitoneal adenopathies; aortic wall without aneurysmal dilation with a fusiform lesion of well-defined borders with homogeneous soft tissue density surrounding the aorta and the cava. To better characterize the tomographic findings, an abdominal magnetic resonance imaging with contrast was performed, which is shown in Figure 1.

The patient did not continue the follow-up of the lesions evidenced by imaging and in February 2018 entered again to the emergency department due to abdominal pain in the right flank and iliac fossa radiated to the lumbar region, with an ultrasound report of appendicitis, an appendectomy was performed and because there was not improvement in pain, it was taken again a simple abdominal tomography that showed lobulated masses with infrarenal periaortic soft tissue density until the bifurcation and primitive iliac arteries up to 28 mm thick that conditioned severe compressive effect on the proximal ureters and grade III hydronephrosis; paraclinical tests were taken showing elevation of nitrogen compounds as evidenced in Table 1, with a requirement to perform derivative nephrostomies. Finally, it was taken a biopsy by laparotomy of the retroperitoneal lesions that are shown in Figures 2 and 3.

Table 1. Results of paraclinical tests.

Paraclinical	2016	Feb-18	After nephrostomies (September 06 2018)
Creatinine mg/dl	1,5	6,74	1,8
BUN mg/dl		62	23
GFR (CKD-EPI) ml/min/1.73 m2	53,09	8,8	42.9

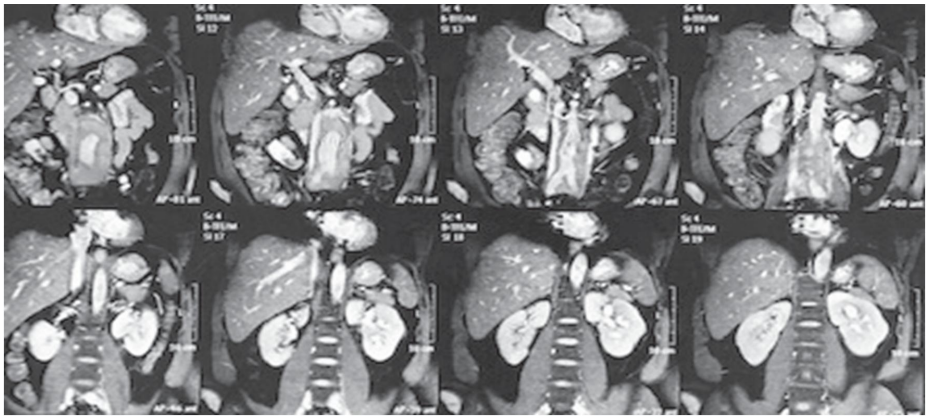

Figure 1. Abdominal magnetic resonance with contrast.

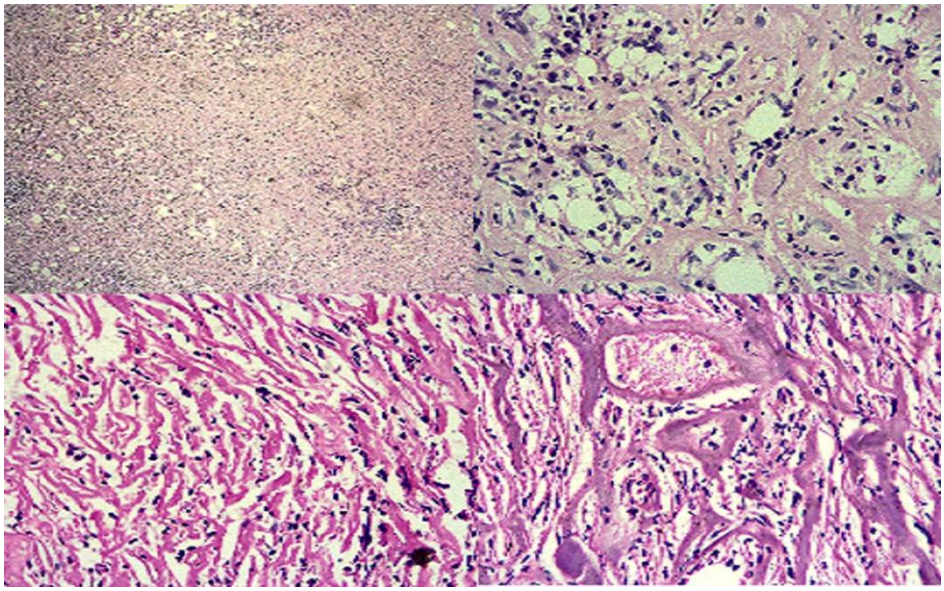

Figure 2. Hematoxylin/eosin. Soft tissues with proliferation of reactive fibroblasts and myofibroblasts with presence of inflammatory lymphoplasmocytic infiltrate, some histiocytes and vascular proliferation are observed. Hematoxylin eosin. A.4x, B-C and D. 20X.

In a medical-surgical meeting it was decided to start prednisolone at 1 mg/kg day, a 6-week follow-up was carried out with significant improvement of urinary volumes and decrease of the nitrogen compounds, continuing in outpatient control by the nephrology service.

Mass up to 20 mm thick that surrounds the abdominal aorta in its infrarenal portion until the aortoiliac division, producing slight displacement of the inferior vena cava and presenting an intermediate signal in T1 and T2, the infrarenal abdominal aorta presents ectasia of up to 28 mm in diameter.

Discussion

Idiopathic retroperitoneal fibrosis (IRF) is a rare entity, characterized by the presence of fibrous

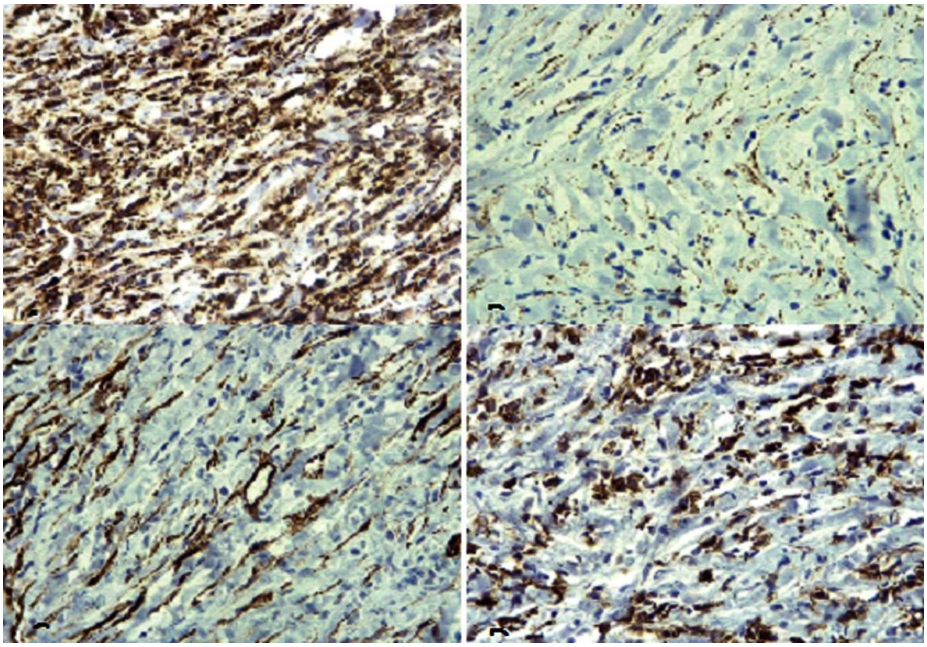

Figure 3. Immunohistochemical studies. **A.** CD 68 that highlights the population of histiocytes. **B.** Beta-catenin that is negative. **C.** CD34 which is positive in vascular walls. **D.** CD45 positive in the population of reactive lymphocytes.

inflammatory retroperitoneal tissue that usually covers the ureters and other abdominal organs, predominantly affecting men with a male to female ratio of 2: 1 ratio, between 50 and 60 years of age,[3] as is the case of our patient.

This pathology is included within chronic periaortitis along with the inflammatory aneurysms of the abdominal aorta, which have no ureteral involvement, while perianeurysmal retroperitoneal fibrosis, as well as IRF has ureteral involvement. IRF is the no n-aneurysmal form of chronic periaortitis.[4]

This entity may be either secondary to some drugs such as ergotamine derivatives: bromocriptine, methysergide, beta blockers, methyldopa, hydralazine; malignant diseases such as Hodgkin and non-Hodgkin lymphoma, sarcomas, colorectal, breast, prostate and bladder carcinoma,[5-7] or idiopathic; the latter in more than 75% of cases.[8]

In the last decade, the concept of IgG 4-related diseases has emerged, encompassing a spectrum of fibro-inflammatory diseases that affect different structures, such as the pancreas, the biliary tract and the lymph nodes.[9]

Histopathologically, it is characterized by lymphoplasmacytic inflammation, pronounced fibrosis and IgG4 plasma cell infiltration.[9]

The cardinal symptom is low back pain, which occurs in approximately 90% of cases,[9] accompanied by nonspecific and heterogeneous symptoms (nausea, emesis, asthenia, adynamia, weight loss, edema in lower limbs, erectile dysfunction) causing a silent evolution of the disease, which in advanced stages can lead to deterioration of renal function of postrenal origin.[10]

The deterioration of renal function evidenced in our patient was secondary to the ureteral obstruction caused by RF, this being the most frequent and serious complication.[3,8] The recovery and long-term prognosis of kidney disease is associated with timely diagnosis and appropriate treatment.[11]

The radiological images obtained are useful for the diagnosis and evaluation of the response to

treatment, such as computed axial tomography (CAT) which also allows us to assess the extent of fibrosis and rule out the presence of associated tumors.[12] In the CAT scan, fibrosis is represented as a soft tissue mass that surrounds the abdominal aorta and the inferior vena cava and, as in the case of our patient, surrounds the iliac arteries and generates medial deviation and extrinsic compression of the ureters.[13] In addition, in the magnetic resonance imaging (MRI) the coefficient values are useful to differentiate between active and inactive lesions.[14]

Due to the variable appearance of RF, malignant causes cannot be distinguished from non-malignant based only on the radiological findings, so a biopsy of the lesion should be performed, this being the most accurate technique for diagnosis.[3]

The mainstay of treatment to improve renal function is to relieve the obstruction of the ureter and other retroperitoneal structures, either with surgical release (open or laparoscopic ureterolysis) or conservative management (double J catheter, nephrostomies) followed by medical management.[15] The possibility of improvement in renal function with these interventions is more than 90%;[16] as in this case where renal function improvement was observed after nephrostomies and starting of corticosteroid therapy (creatinine from 6.74 to 1.8 mg/dl post-nephrostomy).

Other goals of treatment are: stopping the progression of the fibrotic process, controlling systemic manifestations and avoiding relapses or recurrences.[13]

Glucocorticoids are the first line of medical management since they have demonstrated a favorable response and remission in 75 to 90% of patients;[14] among corticosteroids, the most commonly used is prednisolone; the initial dose is 0.75 - 1 mg/kg/day with gradual decrease until reaching 5 to 7.5 mg/day in the next 6 to 9 months.[17-19] It is important to monitor in these patients the metabolic and skeletal effects of chronic use of corticosteroids.

Today, alternative immunosuppressive drugs are also used in combination with corticosteroids in severe cases or as monotherapy when there is resistance to corticosteroids.[20] Azathioprine, mycophenolate, tamoxifen, cyclophosphamide and rituximab are among the options.[8,18,21,22-24]

The duration of treatment of this pathology has not yet been clearly defined, however, some authors recommend 1 to 3 years.[17,19] Large studies and recommendations still remain to be done to be able to establish the best management.

Through this case report, we present the clinical impact that this disease can have, recognizing that it is a pathology that requires a multidisciplinary management (nephrology, rheumatology, urology, general surgery, pathology) with the objective of establishing an early diagnosis and timely treatment that avoids potentially irreversible complications.

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Ethical responsibilities

Protection of people and animals

The authors declare that no experiments were performed on human beings or animals for this research.

Data confidentiality

The authors declare that they have followed the protocols of their workplace on the publication of patient data.

Right to privacy and informed consent

The authors declare that patient data do not appear in this article.

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