

## Unilateral Lumbar Ureteral Obstruction in Idiopathic Retroperitoneal Fibrosis (Ormond's Disease)

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**Abstract :** Idiopathic retroperitoneal fibrosis (RPF) or Ormond's disease is a rare chronic inflammatory disease with an estimated prevalence of 0.2-1.4/100,000 inhabitants and male predominance. This disease is often little known and underdiagnosed by health professionals.

Ureteral involvement is common in this disease and dominates its prognosis with the potential risk of hydronephrosis and renal failure.

It is classically bilateral and predominates in the pelvic ureter. Unilateral ureteral obstruction is much more rare and unusual during RPF.

We report the original observation of unilateral lumbar ureteral obstruction caused by Idiopathic RPF in a Tunisian man.

**Keywords:** Ureteral obstruction, idiopathic retro péritonéal fibroses, Ormond's disease, hydronephrosis

### 1. INTRODUCTION

Described for the first time in 1905 by Albarran and recognized as a separate clinical-pathological entity in 1945 by Ormand, idiopathic or primary retroperitoneal fibrosis (RPF) is a chronic inflammatory disease which is characterized by the encasement of the retroperitoneal structures by a fibro sclerotic process [1, 2].

It is a rare disease, often little known, and often underdiagnosed by health professionals [2-4]. Its prevalence is estimated at 0.2-1.4/100,000 inhabitants [1, 2] with a clear male predominance: sex ratio at 3M/1F [4].

Ureteral involvement is the most common complication of this disease [1]. It is classically bilateral and predominates in the pelvic ureter [1, 2, 4, and 5]. Unilateral ureteral obstruction is much more rare and unusual in this disease [3, 4, and 6].

We report the original observation of unilateral lumbar ureteral obstruction caused by Idiopathic RPF in a Tunisian man.

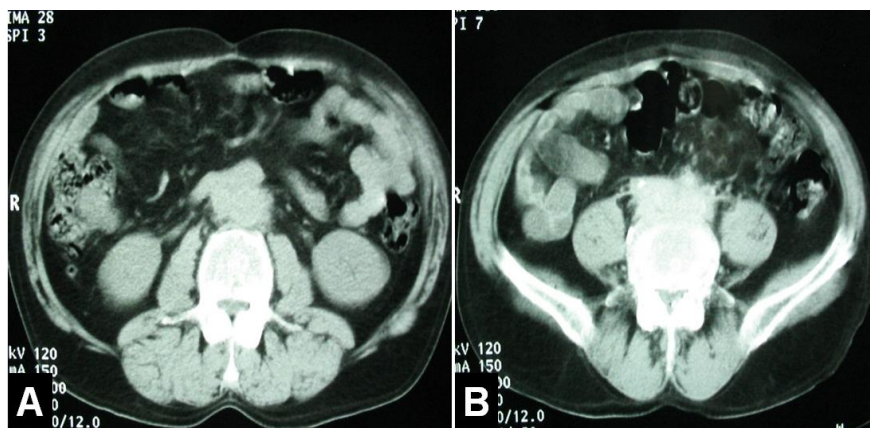
### 2. CASE REPORT

A 56-year-old Tunisian man, without pathological medical history, was explored for chronic pain of the left flank. This pain had

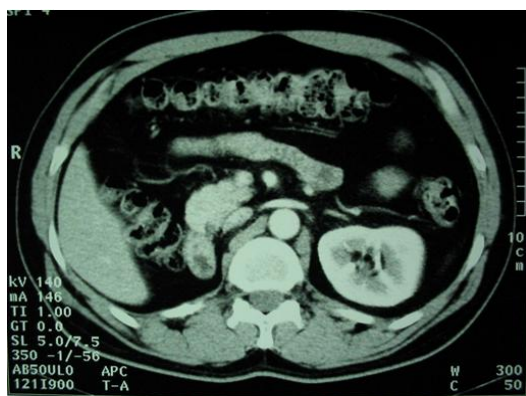
gradually increased in intensity over the past three months, with no fever, lower urinary tract symptoms, or the notion of local trauma.

The somatic examination noted an apyretic patient with conserved neurological, cardiac, and respiratory statuses. The abdomen was slightly painful on palpation without distension. The examination of the left flank noted a positive lumbar contact with perception of a large painful kidney. Basic biology tests were without abnormalities: total blood count, creatinine, calcemia, fasting glycemia, transaminases, lipid parameters, muscular enzymes, uric acid, plasma ionogram, erythrocyte sedimentation rate, C-reactive protein, thyroid tests, and electrophoresis of serum proteins. Urinalysis and urine culture were negative.

Abdominal ultrasound objectified dilated left urinary cavities without visualization of obstacle. Computed tomography confirmed the left hydronephrosis with ureterial sheathing by a fibrous process (Figures 1 and 2) and the existence of a circumferential periaortic fibrous mass (Figure 3). The urographic images showed a left unilateral lumbar ureteral stenosis at the level of the fourth lumbar vertebra with medial ureteral deviation and left pyelocalicel cavities dilation (Figure 4).



**Figure1:** Abdominal CT without (A) and with (B) contrast injection: homogeneous fibrous process in the retroperitoneal space, entrapping the aorta and the left ureter.



**Figure2:** Abdominal CT with contrast injection: left hydronephrosis.



**Figure3:** Abdominal CT-scan with contrast injection: circumferential fibrous process of the abdominal aorta.



**Figure4:** Intravenous urography: unilateral obstruction of the left lumbar ureter with medial deviation, and left pyelocaliceal cavities dilation.

The chest X-ray, electrocardiogram, cardiac ultrasound, and chest CT-scan were free of abnormalities. Histological examination of a laparoscopic biopsy showed nonspecific fibro inflammatory tissue, with no signs of malignancy, micro thrombosis, or vacuities.

The etiological investigations had eliminated secondary RPF (medications, connective tissue diseases, systemic vacuities, cancers, autoimmune diseases, auto inflammatory

diseases...). The diagnosis of idiopathic RPF complicated by left lumbar ureteral obstruction was retained.

The patient was treated with systemic glucocorticoids at 1mg/kg/d for six weeks followed by gradual taper. The evolution was favorable with a progressive resolution of the inflammatory process on the control CT scans. CT scan showed normal appearance of the urinary tract with complete disappearance of the

retroperitoneal fibrous process after eight months.

### 3. DISCUSSION

Kidney complications of RPF are dominated by ureteral obstruction; other rarer types of renal involvement such as renal arteries and veins stenosis, renal atrophy, and different types of associated glomerulonephritis can be cited [1].

Ureteral involvement dominates the prognosis of this disease with the potential risk of hydronephrosis and renal failure requiring early diagnosis and appropriate management without delay [7, 8].

Unilateral ureteral obstruction caused by Ormond's disease remains rare and unusual with only a few sporadic cases reported [3, 4,9]. Indeed, five of the six patients with retroperitoneal fibrosis of Scheel PJ Jr et al, had bilateral ureteral obstruction against only one case of unilateral involvement [10]. Similarly, in Li KP et al series of 37 patients with RPF and ureteral obstruction, the involvement was bilateral in 29 cases against only 8 cases of unilateral involvement [2].

Urgent treatment in these situations is based on urinary diversion to preserve renal function [4, 7]. Conventionally, the disease responds favorably to systemic corticosteroids alone [4, 6, 7] or associated with immunosuppressive agents [1, 2, 10]. Surgical treatment is limited to refractory forms [7].

### 4. CONCLUSION

As rare as it is, idiopathic RPF deserves to be known by healthcare professionals, especially those in primary care. This improved knowledge will make it possible to make the diagnosis in time and avoid the delay in treatment. Unilateral ureteral obstruction remains an unusual and original presentation of Ormond's disease.

### REFERENCES

- [1] Vaglio A, Maritati F. Idiopathic Retroperitoneal Fibrosis. *J Am SocNephrol*. 2016; 27(7):1880-9.
- [2] Li KP, Zhu J, and Zhang JL, Huang F. Idiopathic retroperitoneal fibrosis (RPF): clinical features of 61 cases and literature review. *Clin Rheumatol*. 2011;30(5):601-5.
- [3] Yan T, Wang Y, Liu Z, Zhang X, Wu Q, Xi M. Idiopathic retroperitoneal fibrosis causing unilateral ureteral and sigmoid colon obstruction: A case report. *Medicine (Baltimore)*. 2017; 96(7):e6105.
- [4] Ghieh D, Saade C, El-Merhi F. An atypical manifestation of idiopathic retroperitoneal fibrosis: A case report. *Urologia*. 2020 Feb 5:391560319899302. doi: 10.1177/0391560319899302. [Epub ahead of print]
- [5] Sandhu A, Brickner L, Chen M. Ureteral Strangulation by Fibrosis: A Cold Case Report of Ormond's Disease. *Case Rep Nephrol*. 2011; 2011:302963.
- [6] Tan WP, Hwang T, Medeiros R, Pessis DA. An Atypical Presentation of Retroperitoneal Fibrosis. *Curr Urol*. 2017; 10(3):157-159.
- [7] Nelius T, Reiher F, Lindenmeir T, Kalinski T, Rau O, Filleur S, et al. Idiopathic retroperitoneal fibrosis (Ormond'sdisease). *AktuelleUrol*. 2006;37(4): 284-8.
- [8] Hadzi-Djokić J, Pejčić T, Bašić D, Vukomanović I, Dzamić Z, Aćimović M, et al. Idionathic retroneritoneal fibrosis: A Report on 15 Patients. *Vojnosanit Pregl*. 2015; 72(10):92 8-31.
- [9] Cornud F, Blangy S, Sibert A, Benacerraf R. Abdominal echography in the diagnosis and monitoring of retroperitoneal fibrosis. Apropos of 2 cases. *J Radiol*. 1983;64(2):111-5.
- [10] Scheel PJ Jr, Piccini J, Rahman MH, Lawler L, Jarrett T. Combined prednisone and mycophenolatemofetil treatment for retroperitoneal fibrosis. *J Urol*. 2007; 178(1):1 40-3.

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