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Spontaneous Remission of Idiopathic Retroperitoneal Fibrosis: A Case Report

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♦ Introduction:

Retroperitoneal fibrosis (RPF) or initially called Ormond's disease [1] is a rare disease characterized by the presence of inflammatory and fibrous tissue in the retro peritoneum. The tissue is generally localized around infrarenal portion of the abdominal aorta and iliac arteries, often encasing the ureters or other abdominal organs [2].
We report a case of idiopathic retroperitoneal fibrosis associated with left hydronephrosis.

♦ Abstract :

Retro péritonéal fibrosis is a rare disease characterized by the presence of inflammatory and fibrous tissue in the retro peritoneum. it 's generally localized around infrarenal portion of abdominal aorta and iliac arteries, often encasing the ureters or other abdominal organs .
Such a spontaneous regression of fibrosis is rare and is not well described in the literature; our aim is to clarify the possible Spontaneous remission of this disease.
We describe the case of an 54-year-old woman suffered from a moderate lumbar pain which appeared one year ago, without other associated urinary signs, and discuss the management of retroperitoneal fibrosis.
Key-words: Retro péritonéal ,fibrosis , idiopathic , regression,spontaneous

♦ Case History

A 54-year-old woman, mother of 3 children, with a history of pulmonary tuberculosis treated for six months with antibacillaries.
The patient suffered from a moderate lumbar pain which appeared one year ago, without any other associated urinary signs.
The physical examination was normal; particurarly absence clinical signs of infection or renal failure, the patient had normal value of hemogram parameters ; CRP at 9 mg/l, the level of blood creatinine was high as 15 mg/l).
The cytobacteriological examination of the urine (CBEU) was sterile, the reno-bladder ultrasound shows a left ureteropyelocaliciel dilatation. The Uroscan showed a retro-peritoneal tissue thikning encasing the aorta below the renal arteries extanded to internal and external iliac arteries, this retroperitoneal mass is slightly enhanced after injection of the iodine contrast product. It encases also the left ureter in its initial part, the inferior vena cava and the left renal vein. With a decrease of secretion and excretion of the left kidney(figure 1).
the patient underwent a bilateral ureteral stenting in the operating room .
After the stenting procedure, the lumbar pain resolved, and renal function was restored .
Computed tomography scan was performed after six months of loss of sight and after stent placement, had showed marked improvement of the retroperitoneal fibrosis, with a maximum
thickness estimated at 7mm at the distal part of the aorta . Based on the CT findings, the decision was made to continue to observe the patient and not initiate medical therapy and the bilateral ureteral stent was changed.
Another CT scan had showed a total regression of retroperitoneal fibrosis with a hypotrophic left kidney without pyelocaliceal dilatation and the bilateral ureteral stenting in place (figure 2).

♦ Discussion:

The primary goal of treatment is preserving renal function. For that reason, idiopathic RPF with absence of ureteral obstruction requires neither medical or urological treatment but only active surveillance.

Generally, in idiopathic RPF causing ureteral obstruction, a good outcome for renal function is expected, although long-term steroid therapy along with urinary drainage is often required.[3] From the point of view of urologists, the final goal of treatment should be resolution of ureteric obstruction requiring drainage and withdrawal of steroid therapy.

As aforementioned, the first-line treatment is glucocorticoid therapy, although the protocol is not standardized. The treatment is usually initiated with 0.6–1 mg/kg/day of PSL for 2– 4 weeks; then the dosage is gradually tapered and maintained with 2.5–5 mg/day for >6 months.[3].

Ureteral obstruction is usually managed with conservative procedures, such as ureteral stenting or percutaneous nephrostomy(PNS). However, both ureteral stents and PNS are harmful. In addition to the necessity of regular exchange, ureteral stents are frequently associated with bladder irritability, hematuria, encrustation and urinary tract infection[4],whereas PNS is associated with impaired QOL and catheterrelated problems, including infection, obstruction and dislodgement. So we should attempt to remove the ureteral stent or PNS as soon as possible, because ureteral obstruction can be resolved even during planned administration of a glucocorticoid.

Retrograde or antegrade pyelography should be carried out to confirm the patency of the ureter at the time of exchange of the stent or PNS tube. If resolution of ureteral obstruction is confirmed, the ureteral stent or PNS can be removed.

In patients who can not obtain such a condition through medical treatment, invasive surgical treatment by Ureterolysis should be considered.

spontaneous regression of idiopathic retroperitoneal fibrosis is very rare, and there are four cases reported in the literature[5-8] , By our knowledge, there are not cases of reactivation following spontaneous regression have been published .

- **CONCLUSION:**

Idiopathic RPF is highly sensitive to glucocorticoid therapy, and the prognosis for renal function is relatively good.

in cases where there is a contre indication to glucocorticoids, spontaneous resolution of idiopathic retroperitoneal fibrosis is possible in selected cases, even if the data in the literature are rare.

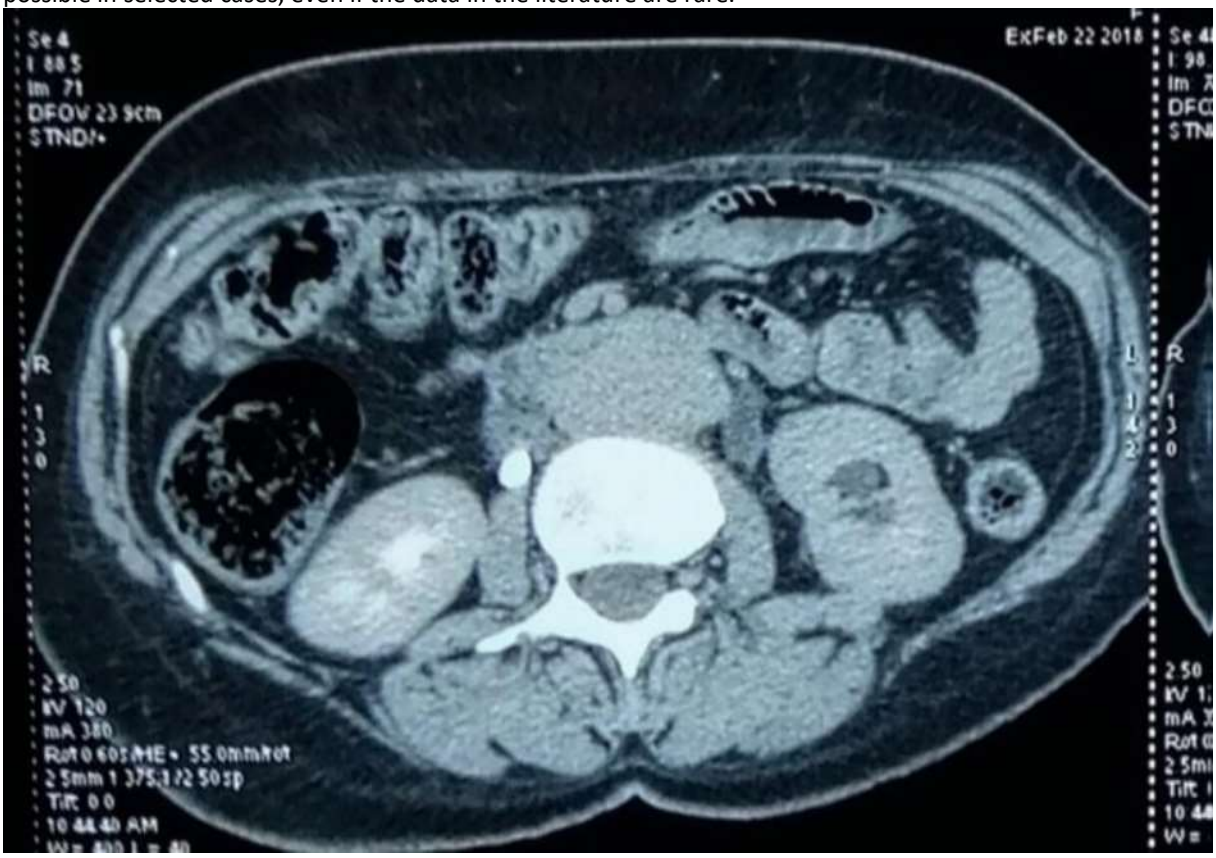

Figure 1: CT shows a retroperitoneal tissue processus incasing the aorta, the vena cava and the left ureter with a ureterohydronephrosis.

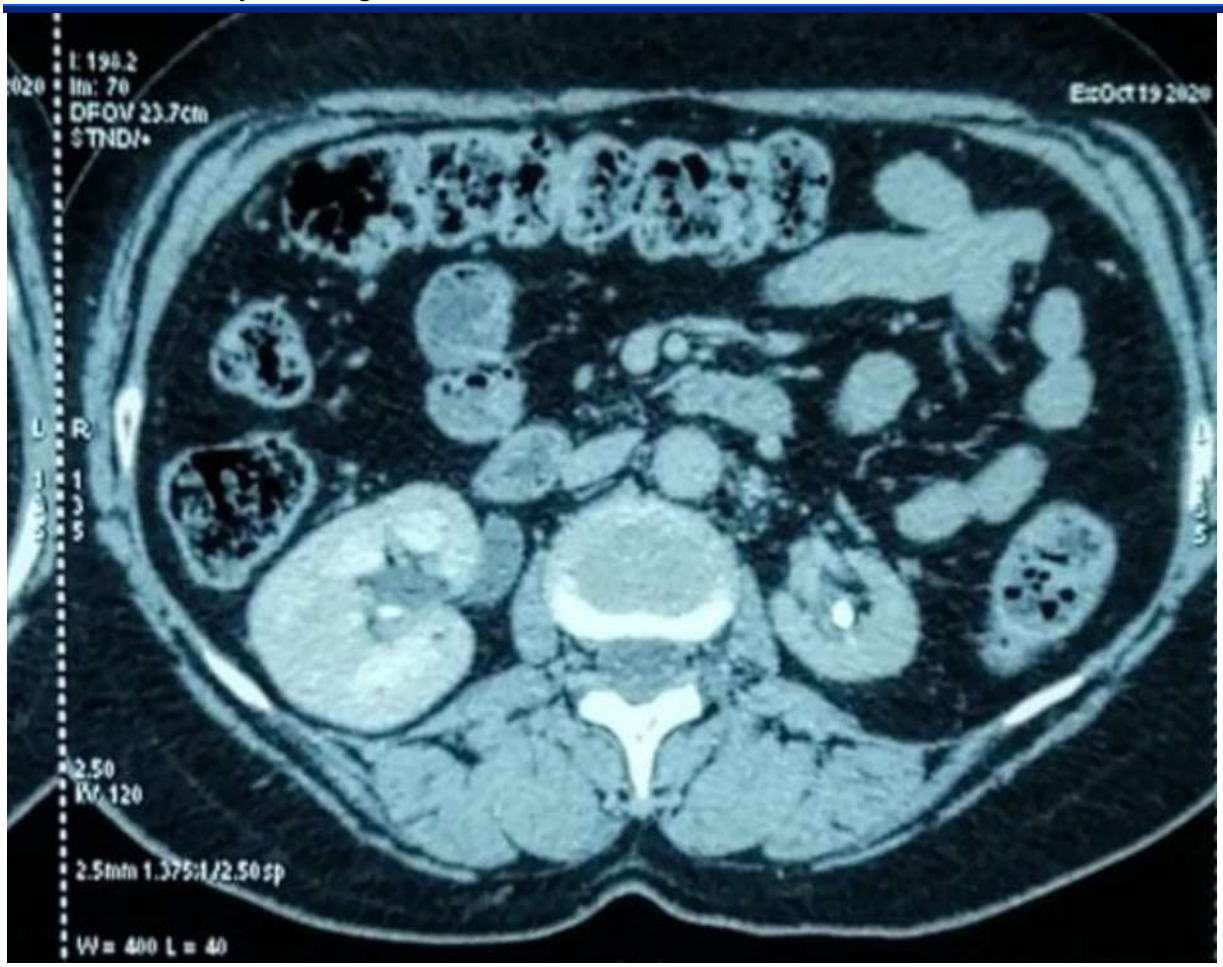

Figure 2: CT shows total regression of RPF with an hypotrophic left kidney .

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