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Case Report

Retro-Peritoneal Fibrosis Refractory to Corticotherapy: Case Report

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Abstract

This is a case report Retroperitoneal Fibrosis where the authors explore an alternative method of treatment that consists of surgical exploration with apposition of a flap around the ureter. Although this is a quite rare disease where clinical manipulation normally is the first option, surgical repair may represent an attractive option of treatment.

Keywords: *Fibrosis; Retro-Peritoneal; Ureterolysis; Flap; Laparotomy*

Introduction

Retroperitoneal fibrosis (RPF) is a rare disease characterized by the presence of fibro-inflammatory tissue that often forms in the sub-renal portion of the abdominal aorta and iliac arteries. It frequently invades surrounding structures: the ureters and the inferior vena cava. It is idiopathic in two-thirds of cases; the remaining third may be secondary to infections, abdominal surgery, taking drugs or malignancies. Recently, idiopathic RPF is reported with the manifestations of IgG4 disease. Surgical treatment consists of bilateral ureterolysis with flap. We present case of retro-peritoneal fibrosis resistant to corticosteroids, treated by laparotomy, bilateral ureterolysis with flap.

Case Representation

Mr. HN, 52 years old, with a history of HIV infection with triple therapy, hepatitis B treated and cured, appendicectomy, repair of bilateral inguinal hernia, diabetic. The patient had bilateral low back pain without fever which had pushed him to the emergency. Biological assessment showing acute renal failure with serum creatinine at 65.45 mg/l (5 - 13 mg/l); 576 µmol/l (50 - 110 µmol/l) and serum potassium at 5.8 mmol/l (3.5 - 5 mmol/l). Abdominopelvic CT scan showing retro-peritoneal fibrosis that invades the large vessels and ureters resulting in moderate bilateral pyelocalyceal dilatation and increased right (Figure 1). Initial management consisted of an internal urinary drainage by ureteral double J tube. The evolution was marked by the normalization of renal function with control creatinemia one week later at 11.14 mg/l (98 umol/l) and serum potassium returned to 3.8 mmol/l. The patient was treated with steroids with change of the double J tube every six (06) months. The TDM control shows persistence sub Aortic retro-peritoneal fibrosis, appearing stable at 20 mm of thickness by 40 mm width. Abdominal aorta and common iliac arteries remain of usual and permeable caliber (Figure 2). The change of double J tube with the persistence of ureteral obstruction which was diagnosed on retrograde uretero pyelography (RUP) was considered therapeutic failure to corticosteroid therapy after two years of treatment. For this clinical situation, the surgery was indicated. Laparotomy by median incision and entering to the peritoneal cavity, we found epiploic adhesions at the right iliac fossa due to the ap-

pendicectomy which was released. We dissect the left colon for release the left ureter whose wall is very thick. His liberation was difficult at the iliac cross.

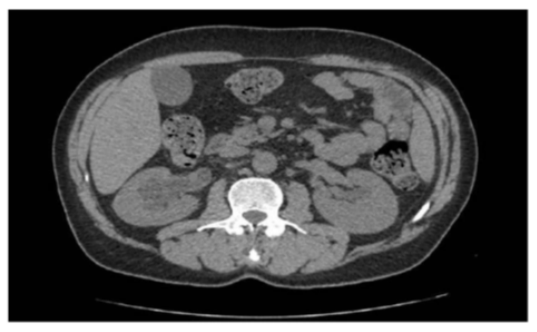

Figure 1: *CT-scan: Retro-peritoneal fibrosis sheathing the vessels and ureters.*

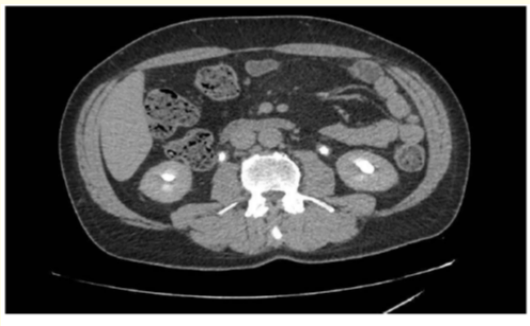

Figure 2: *T-scan: Retro-peritoneal fibrosis sheathing the vessels and ureters. Control scan persistence of Retro-peritoneal fibrosis resist to corticosteroid therapy.*

We mobilized the lumbar ureter. Down, the ureter is released as low as possible (Figure 3): but the area is less fibrous after passing the iliac cross.

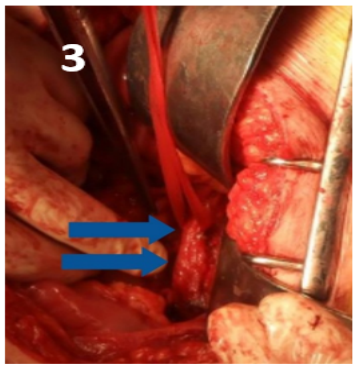

Figure 3: *Ureterolysis to the pelvic part blue arrows: ureter.*

Biopsy's Samples were taken for anatomopathological analysis, whose response was in favor of inflammatory tissue without malignancy observed.

Preparation of large peritoneal flap with a low inferior implantation was taken (Figure 4): positioning between the released ureter and the deep plane by fixing the peritoneum with 3/0 vicryl points (Figure 5).

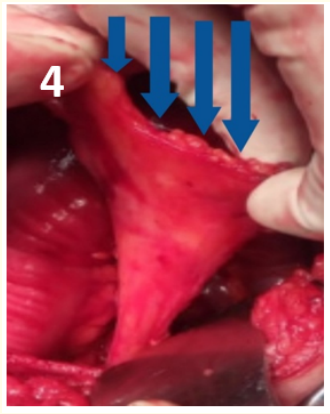

Figure 4: *The arrows represent the garden flap.*

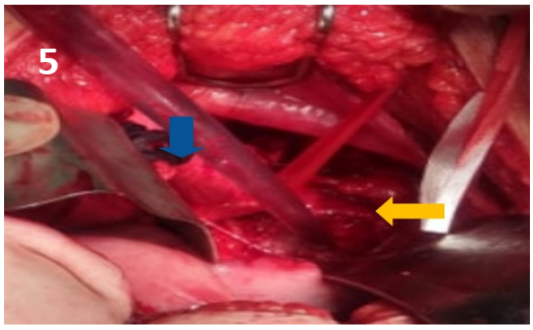

Figure 5: *Flap interposition: flap of the garden (blue arrow), ureter (yellow arrow).*

On the right side, the release of the ureter is also difficult but it is also performed on the finger, without vascular wound. we could not recover a peritoneal flap: we then interposed the large omentum which descended without tension which was fixed to the vicryl 3/0. The postoperative consequences were simple. The patient discharged. Double J tube were removed at the 8th week.

We present through this clinical illustration and review of the literature, the treatment with open surgery of the Retro-peritoneal fibrosis.

Discussion

Retro peritoneal fibrosis (RPF) is a disease whose pathogenesis is still obscure and is considered idiopathic disease. In 1980 Mitshinson and Parums considered retro-peritoneal fibrosis as a local inflammatory reaction exacerbated by aortic atherosclerosis. This reaction is triggered against the lipids contained in the plaque of atherosclerosis such as oxidized LDL and ceroid [1].

However, patients with retro-peritoneal fibrosis often have general symptoms, elevated levels of inflammation markers, anti-body positive, related autoimmune diseases, involvement of other organs, and good response to immunosuppressants. These results suggest that Retro peritoneal fibrosis (RPF) is a manifestation of systemic disease rather than an exacerbated local reaction to atherosclerosis [2].

Some studies have suggested that the origin of Retro peritoneal fibrosis (RPF) would be primary aortitis, which subsequently provoked a peri-aortic fibro inflammatory response. Vascular inflammation can then affect the vascular segments including the thoracic aorta and its branches, the gastrointestinal arteries and predominate at the level of the adventitia; vasculitis of vasa varum can also be observed [3].

Recently, some study have suggested that idiopathic Retro peritoneal fibrosis (RPF) is part of systemic inflammatory condition known as IgG4 [4]. It has been identified in 50% of patients who have already had a diagnosis of idiopathic Retro peritoneal fibrosis (RPF) [5].

The clinical signs of the disease are nonspecific, highly variable and related to the mechanical effect of Retro peritoneal fibrosis (RPF) on nearby structures: such as abdominal pain, low back pain like our patient, renal colic, limb edema, deep vein thrombosis, intermittent claudication of lower limbs, testicular pain, hydrocele [6].

Biologically, impaired renal function may be indicative of Retro peritoneal fibrosis (RPF) [7]. This alteration of renal function varies according to degree of obstruction, CRP is often increased in 69% of cases [8].

Presence of low back pain with impaired renal function in biology, scan is mandatory to evaluate the upper urinary tract, including an ultrasound and more specifically a uroscanner looking for obstacle in the urinary tract [9]. The typical appearance is a well-defined, irregular mass of periaortic tissue that extends from the level of the iliac vessels and progresses in the peritoneum by enveloping the ureters and inferior vena cava (IVC) [10].

The treatment of idiopathic Retro peritoneal fibrosis (RPF) [11] remains empirical and modeled on the management of autoimmune or inflammatory diseases.

Anecdotal cases of spontaneous resolution have been reported, and some asymptomatic patients with no impact on adjacent structures could be survived only.

For symptomatic patients, corticosteroid therapy remains the first-line of treatment, but there is no consensus regarding the dose (0.5 - 1 mg/kg/day) and duration. The most reported protocols in the literature are:

- 60 mg/day for two months (1J/2), then progressive decrease to 5 mg in two months, then 5 mg/day for two years.

This is the most used protocol

- 40 - 60 mg/day for six weeks then gradual decrease over 2-3 months to reach 5 - 10 mg. Duration: 6 - 12 months.
- 1 mg/day (max 80 mg) for four weeks, then progressive decrease to reach 10 mg/day in 2 - 3 months. Duration: 12 - 18 months.

The authors concluded that corticosteroids have a rapid effect on clinical symptoms, biological parameters and, more rarely, radiological images, which makes them a good induction therapy. However, their long-term use resulted in more adverse effects than tamoxifen.

Tamoxifen, by its anti-fibrotic properties, can be used in relays of corticosteroids for a cortisonic saving. Only one prospective trial comparing the efficacy of prednisone against tamoxifen in preventing recurrence has been conducted. This study showed that tamoxifen is less effective than prednisone in the presence of recurrence.

Other cortisone sparing strategies have been tried with mycophenolate mofetil, cyclophosphamide and azathioprine. Strategies combining prednisone and mycophenolate mofetil or azathioprine have been successful. On the other hand, the side effects in the largest study combining cyclophosphamide and corticosteroids are probably unacceptable.

Although various immunosuppressive agents have been successfully used to treat Retro peritoneal fibrosis (RPF), none of them have been systematically studied in relapsed patients. Recently, study has shown that the combination of methotrexate and prednisone is a viable option for treating recurrent Retro peritoneal fibrosis (RPF). This diet has brought remission of significant proportion of patients with normalisation of inflammatory markers, excellent renal function and limited toxic effects. After stopping treatment, patients may be at high risk of relapse and should be closely followed. More recently, biotherapies have also been used in the treatment of resistant Retro peritoneal fibrosis (RPF). These agents may have a role, but more important studies are needed.

Surgically, in the case of dilation of the urinary tract, double-J tube or percutaneous nephrostomy is required during the acute phase in combination with drug therapy. Permanent ureteral stents are the most common form of drainage; they are usually removed after 3 months of treatment.

Once the disease is stabilized by the drug treatment and if the patent urinary obstruction persists, the surgery can be proposed. It allows ureterolysis with deep biopsies of the Retro peritoneal fibrosis, then protection of the ureters by lateral repositioning with intra-peritonealization and/or use of omental or peritoneal flap, required to restore renal function.

The surgical approach can be performed either in the open laparotomy or laparoscopic with or without robot assistance. Minimally invasive techniques have a complication rate similar to the conventional method and are also effective. In addition, they may provide shorter convalescence and less use of postoperative analgesia and blood transfusion.

However, surgical procedures may be associated with significant risks of complications, including ureteral wounds, ureteral devascularization and stenosis, urinary fistula, recurrent fibrosis and thromboembolic complications.

Conclusion

Retro peritoneal fibrosis (RPF) is a rare disease, most often idiopathic. The diagnostic procedure is not codified. Despite the lack of consensus, corticosteroids associated with urinary drainage in case of dilation of the urinary tract would be the first-line treatment. Surgical treatment with ureterolysis with flap, is one of the promising therapeutic options.

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