SHORT COMMUNICATION

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Idiopathic retroperitoneal fibrosis: A report on 15 patients

Idiopatska retroperitonealna fibroza: rezultati lečenja 15 bolesnika

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Abstract

Background/Aim. Retroperitoneal fibrosis (RPF) represents a chronic pathological process characterized by fibrosis which entraps and compresses the ureters and the great blood vessels in the retroperitoneal space. A specific form of RPF is idiopathic RPF, an uncommon collagen vascular disease of unclear etiology. The series of 15 patients which underwent open surgical repair due to idiopathic RPF is presented herein. **Methods.** From 1989 to 2012, 11 male and 4 female patients underwent surgery due to primary RPF. The ureters were entrapped unilaterally (7 patients), or bilaterally (8 patients). Major symptoms included low back pain due to hydronephrosis (9 patients), uremia (4 patients), and urinary tract infection (2 patients). The diagnosis was based on intravenous urography (IVU), retrograde ureteropyelography and computed tomography (CT). **Results.** Surgical procedures included intraperitoneal ureteral displacement (8 patients) and ureteral wrapping with omental flap (6 patients). One patient underwent bilateral ureteral stenotic segments resection and oblique ureterography, followed by wrapping with omental flap. Pathological examination confirmed primary RPF in all patients. The mean operative time was 3.5 h (range 2.5–4.5 h). The average intrahospital stay was 21 days (range 16–26 days). The mean follow up was 32 months (6–46 months). During the follow up, 12 patients had improvement on IVU. **Conclusion.** Early recognition of signs and symptoms of RPF is of the utmost importance for the outcome. Surgical procedures, including ureteral wrapping with omental flap, or intraperitoneal ureteral displacement, usually represent definitive treatment.

Key words:
retroperitoneal fibrosis; diagnosis, differential; urologic surgical procedures.

Apstrakt

Uvod/Cilj: Retroperitonealna fibroza (RPF) predstavlja hronični patološki proces koji karakteriše fibroza koja obuhvata i pritiska ureter i velike krvne sudove u retroperitonealnom prostoru. Specifičan oblik RPF je idiopatska RPF, retka vaskularna kolagena bolesti nejasne etiologije. U ovom radu predstavljena je grupa od 15 bolesnika koji su operisani zbog idiopatske RPF. **Metode.** Od 1989. do 2012. ukupno 11 muškaraca i četiri žene operisani su zbog primarne RPF. Ureteri su bili zahvaćeni jednostrano kod sedam bolesnika ili bilateralno kod osam bolesnika. Glavni simptomi bili su lumbalni bol zbog hidronefroze, uremija i infekcije urinarnog trakta. Dijagnoza je postavljena na osnovu intravenske urografije (IVU), retrogradne ureteropijelografije i kompjuterizovane tomografije (KT). **Rezultati.** Primenjene su sledeće hirurške procedure: intraperiteonalno postavljanje uretera kod osam bolesnika i obmotavanje uretera omentalnim flapom kod šest bolesnika. Kod jednog bolesnika učinjena je bilateralna resekcija stenotičnog segmenta i ureterografija, a potom i obmotavanje uretera omentalnim flapom. Patološki pregled potvrdio je postojanje primarne RPF kod svih bolesnika. Prosečno trajanje operacije bilo je 3,5 sata (od 2,5 do 4,5 sata). Prosečni boravak u bolnici bio je 21 dan (od 16 do 26 dana). Prosečno praćenje iznosilo je 32 meseca (od 6 do 46 meseci). Tokom praćenja, kod 12 bolesnika primenom IVU konstantovano je poboljšanje. **Zaključak.** Rano prepoznavanje znakova i simptoma RPF od najveće je važnosti za ishod lečenja. Hirurški zahvati, uključujući obmotavanje uretera omentalnim flapom ili intraperitonealno postavljanje uretera, obično predstavljaju definitivno lečenje.

Ključne reči:
fibroza, retroperitonealna; dijagnoza, diferencijalna; hirurgija, urološka, procedure.

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Introduction

Retroperitoneal fibrosis or Ormond's disease is an uncommon collagen vascular disease of unclear etiology. The disease was documented by Albaran in 1905 for the first time and rediscovered by John Kelso Ormond in 1948 [1, 2].

The RPF is a chronic diffuse retroperitoneal inflammatory process that can entrap the retroperitoneal structures, mainly the ureters and the great vessels. Fibrosis may involve the mediastinum, scrotum and the base of mesentery, as well. The symptoms are nonspecific, including flank pain, malaise, anorexia and renal failure. Some patients are asymptomatic and are diagnosed with RPF during the follow-up of the primary disease [3–5]. The RPF is generally idiopathic, often in the presence of inflammatory abdominal aortic aneurysm or syndrome of vasculitis. However, RPF can appear secondary to the use of certain drugs, malignant diseases, infections, radiotherapy and surgery. Some cases are related to gynecological malignancy.

In cases with renal failure, retrograde ureteropyelography may reveal the length of the involved ureter. In addition, computed tomography (CT) urography, or magnetic resonance imaging (MRI) will help to evaluate the extent of fibrotic changes in the retroperitoneum.

The initial treatment of primary RPF consists of ureteral stenting, followed by immunosuppressive therapy. However, the surgical ureterolysis represents definitive treatment for the vast majority of cases. While open ureterolysis still represents traditional option, laparoscopic ureterolysis is widely accepted today. In the centers where it is available, laparoscopic RPF repair offers additional advantages of shorter hospital stay and reduced transfusion requirements [6]. However, some authors think that the limitations of laparoscopic RPF repair are the cases with very long ureteral entrapment and stricture [7].

The aim of this study was to present patients which subjected to the open surgical repair due to idiopathic RPF.

Methods

From January 1989 to December 2012, a total of 15 patients (11 male and 4 female), with a mean age of 56.4 years, (range 28–72 years), underwent open surgery due to RPF. The patients with intrinsic ureteral obstruction, or another cause for extrinsic obstruction, were excluded from this series.

Preoperative assessment included complete laboratory blood and urine analysis, physical examination, digital rectal/or vaginal examination, abdominal ultrasonography, intravenous urography (IVU) and/or CT-urography. The IVU was performed in order to present the renal excretion, ureteral course, and the site of ureteral obstruction. Recently, CT-urography has been more commonly used, as it provides more details, including the assessment of the retroperitoneal scarring process etc. The biopsies of retroperitoneal tissue were done in all the patients. The patients with impaired renal function or a documented allergy to contrast material underwent retrograde ureteropyelography.

Results

Medial unilateral or bilateral ureteral deviation and proximal dilation seen on IVU are typical for the diagnosis of RPF (Figure 1).

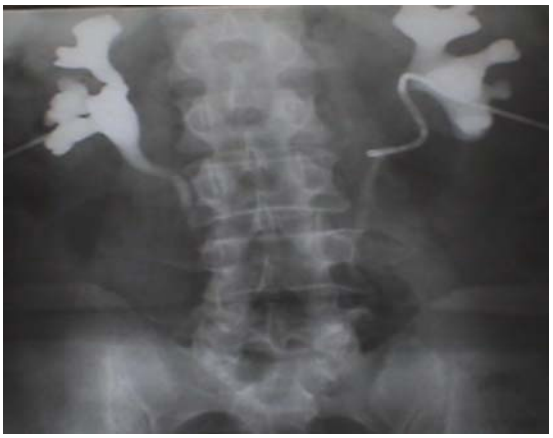

Fig. 1 – Bilateral retroperitoneal fibrosis. Medial deviations of both ureters. Percutaneous nephrostomy tubes placed into both kidneys.

A total of 23 renoureteral units were affected: seven patients (2 female and 5 male) had unilateral, while eight patients (2 female and 6 male) had bilateral ureteral entrapment. The most common symptoms and signs were low back pain due to hydronephrosis (9 patients), uremia (4 patients) and urinary tract infection (2 patients).

The mean time from the appearance of the first symptoms to the presentation was 15.8 months.

Severe hydronephrosis or hydronephrosis associated with impaired renal function were the indications for percutaneous nephrostomy (PCN) in nine renoureteral units; PCN tube was retained until the optimization of renal function. A double-J stent was inserted in 14 renoureteral units preoperatively, in order to facilitate the identification of the ureters during the surgery.

In all the patients, the retroperitoneal space was exposed through the midline laparotomy. There were no perioperative complications (Figure 2).

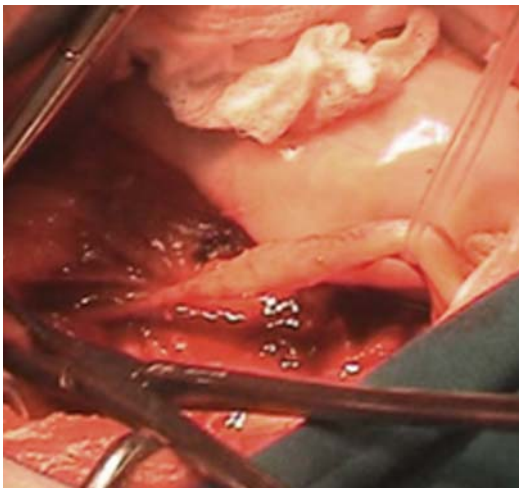

Fig. 2 – The ureter with the stenotic segment deliberated from fibrosis.

After mobilization of the ureters and the dissection of periureteral fibrosis, three different surgical procedures were performed: intraperitoneal ureteral displacement (8 patients), ureteral wrapping with omental flap (6 patients) and bilateral resection of stenotic ureteral segments followed by omental flap wrapping (1 patient) (Figures 3–5).

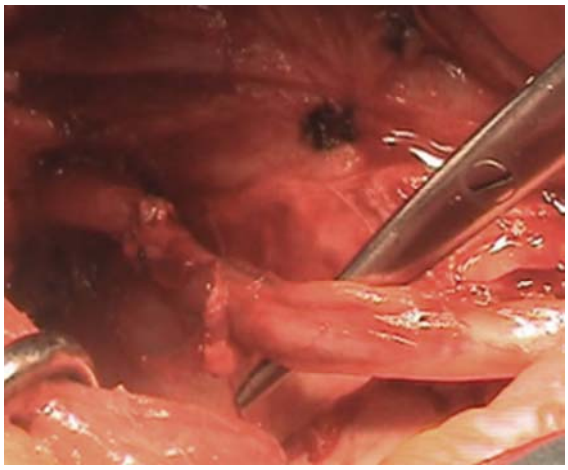

Fig. 3 – Stenotic ureteral segment was removed and the ureter reanastomozed.

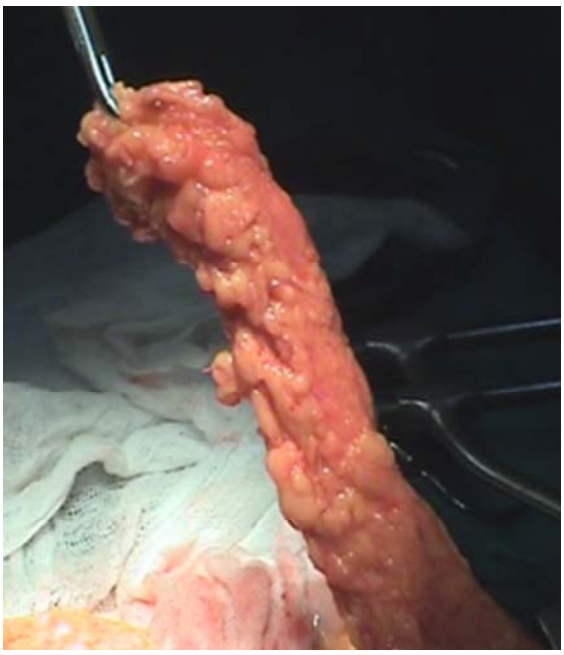

Fig. 4 – Omental flap.

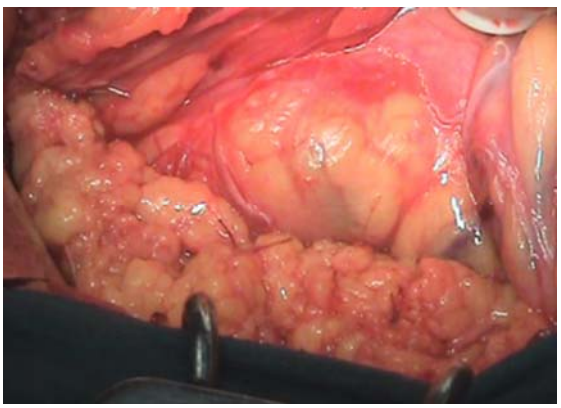

Fig. 5 – The ureter wrapped into the omental flap.

Ureteral stents were removed on the day 21 postoperatively. During the follow-up, serum creatinine measurement, abdominal ultrasonography, IVU and CT-urography were performed.

Pathological examination confirmed primary RPF in all the patients. The mean operative time was 3.5 h (2.5–4.5 h). The average intrahospital stay was 21 days (16–26 days). The mean follow-up was 32 months (6–46 months). During the follow-up, 12 of the patients had improvement on IVU.

Two patients had further deterioration of renal function. One patient had unresolved pelvicalyceal dilation, which required double-J placement.

Discussion

The term retroperitoneal fibrosis denotes the presence of fibroinflammatory tissue in the retroperitoneal space, which surrounds and entraps the great vessels and the ureters.

Primary or idiopathic RPF is probably the result of local inflammatory response to various antigens. The diagnosis of true idiopathic form of RPF is likely in all patients where no potential causative agent may be identified. However, its pathogenesis seems to be related to IgG4 autoimmune mechanisms.

From the other hand, the etiology of secondary forms of RPF is diverse and it can be the consequence of various medications, infections, traumas and malignancies. The RPF secondary to aortic aneurysm is probably the result of inflammatory response induced by leakage of lipids from aneurysm [1, 8]. RPF associated with rheumatoid arthritis or systemic lupus erythematosus probably represents the autoimmune reaction. In addition, these cases usually respond well to steroids and immunosuppressive therapy [9, 10]. Other frequent causes of secondary RPF are previous abdominal or retroperitoneal surgery, retroperitoneal hematoma, and/or extravasation of the urine, and very commonly, radiotherapy. Drugs that potentially cause RPF include methysergide, beta-adrenergic blockers, lysergic acid diethylamide, methyldopa, amphetamines, phenacetin, pergolide, cocaine etc. [1, 11, 12].

The classical clinical signs of RPF are hydronephrosis and medial ureteral deviation seen on IVU or CT-urography. Retrograde ureteropyelography is indicated in patients with impaired renal function; it can be followed by ureteral stenting which facilitates intraoperative identification and handling of the ureters. Percutaneous nephrostomy is indicated in patients with severe hydronephrosis, as a primary treatment before surgery. Preoperative biopsy of retroperitoneal mass is useful to provide information regarding the type of the disease. It is usually performed under the CT guidance, using a true-cut needle, or by fine needle aspiration [13].

Conservative treatment is indicated in patients with RPF associated with some connective tissue disease or with inflammatory abdominal aortic aneurysm. Frequently used medications are methylprednisolon, azathioprine, penicilamine, tamoxifen and various immunosuppressive agents [9, 10, 14]. The last can be used for six months after the surgery, to prevent recurrence [15].

Ureteral stenting or PCN as definitive measures are indicated only in patients with a significant comorbidity.

Surgical management of RPF traditionally included open ureterolysis through the median laparotomy. Rarely, an extensive loss of ureteral length requires various reconstructive procedures, like Boari bladder flap, or even ureteral replacement with the ileum [16, 17].

In the described series, there was no need for ureteral replacement. The most frequent procedure was intraperitoneal ureteral displacement. The procedure starts with ureterolysis, or freeing up the ureters from the fibrous tissue. After that, the ureter can be pulled laterally and completely covered with the folding of the parietal peritoneum. In cases with the abundant omentum, a similar procedure can be performed using the omentum or omental flap. In one patient, there was complicated situation, due to severe fibrosis and stenosis of both ureters. The authors performed bilateral resection of the ureteral segments, oblique end-to-end anastomosis, stenting and the omental flap wrapping.

Recently, more and more RPF repairs have been performed using laparoscopy. Laparoscopic RPF repair is followed by low morbidity; it allows effective ureterolysis and the interposition of omentum or peritoneum. The only possible limitation for laparoscopy is a very long ureteral entrapment and stenosis [18–20].

Conclusion

Retroperitoneal fibrosis represents an uncommon disease of heterogeneous etiology, with the great impact on upper urinary tract. Idiopathic retroperitoneal fibrosis is frequently discovered incidentally, as upper urinary tract obstruction of unknown etiology.

While conservative therapy is rarely successful, surgery still represents the only curative treatment.

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