CASE REVIEW

Robotic Ureterolysis, Retroperitoneal Biopsy, and Omental Wrap for the Treatment of Ureteral Obstruction Due to Idiopathic Retroperitoneal Fibrosis

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Retroperitoneal fibrosis (RPF) is a rare disorder of unclear etiology characterized by chronic inflammation of the retroperitoneum, which can involve any of the retroperitoneal structures, most notably the ureters, aorta, and vena cava. Historically, open biopsy, ureterolysis, and transpositioning or omental wrapping of the involved ureter(s) have been the preferred surgical treatments of RPF, with success rates greater than 90%. More recently, successful laparoscopic biopsy, ureterolysis, and ureteral omental wrapping and intraperitonealization have been described. We report the first case in the literature of idiopathic RPF managed with robotic ureterolysis and laparoscopic omental ureteral wrapping.


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Retroperitoneal fibrosis (RPF) is a rare disorder of unclear etiology first described by the French urologist Albarran in 1906 and then made well known by Ormond’s report in the English literature in 1948. It is characterized by chronic inflammation of the retroperitoneum that can involve any of the retroperitoneal structures, most notably the ureters, aorta, and vena cava. Although its true incidence is unknown, estimates range from one case per 200,000 to 500,000 individuals per year. It occurs predominantly in men in their fifth and sixth decades of life, with a 3:1 male/female ratio, and no ethnic predisposition.
Idiopathic RPF accounts for 60% to 70% of RPF cases.\(^6\) The remainder of cases is due to secondary RPF, which can have any of the following etiologies: malignancy, drug-induced, infectious, traumatic, and autoimmune.

Historically, open biopsy, ureterolysis, and transpositioning or omental wrapping of the involved ureter(s) have been the preferred surgical treatments of RPF, with success rates greater than 90%.\(^7-9\) More recently, successful laparoscopic biopsy, ureterolysis, and ureteral omental wrapping and intraperitonealization have been described.\(^6,10,11\) We report the first case in the literature of idiopathic RPF managed with robotic ureterolysis, retroperitoneal biopsy, and laparoscopic omental ureteral wrapping. We believe that there are several advantages to performing ureterolysis robotically and have begun to perform all of our ureterolysis cases in this manner. Our first case and data are presented here.

**Case Presentation**

The patient was a 48-year-old Caucasian man with hypertension and diabetes mellitus who initially presented in July 2005 to an outside hospital’s emergency department complaining of right flank pain. A computed tomography (CT) scan performed at that time revealed a large mass surrounding the aorta, as well as some moderate hydronephrosis of the right kidney and a delayed right nephrogram. The left ureter appeared uninvolved, and there was no left hydronephrosis.

On April 28, 2006, the patient underwent a successful robotic right ureterolysis with laparoscopic omental wrap. The procedure began with cystoscopy and retrograde pyelography, through the existing stent, to reveal a 2.5-cm narrowing of the mid-ureter, just above the level of the iliac vessels (Figure 2). The right ureteral stent was exchanged, and the patient was re-prepared for surgery and draped in a left modified-flank position for the robotic and laparoscopic portions of the case. A 12-mm trocar and 2 robotic trocars were placed in a wide “V” configuration; 2 assistant trocars (12 mm and 5 mm) were placed in the midline above the camera port (Figure 3).

The da Vinci robotic system (Intuitive Surgical, Sunnyvale, CA) was changed 3 times, and the patient underwent a short course of prednisone, which he was unable to tolerate due to his diabetes mellitus. The mass remained stable in size, according to interval CT scans. In April 2006, the patient presented to us for further evaluation, complaining of right-side flank and back pain that he managed by taking 6 narcotic pain pills per day. He denied any history of dysuria, gross hematuria, fever, nausea, vomiting, dizziness, or headache. He admitted to being an active smoker for the last 30 years.

Physical examination showed the patient to be a well-developed and well-nourished man with a soft, non-tender, and non-distended abdomen, without any masses. There was no costovertebral angle tenderness. Results from the rest of his examination and laboratory workup were unremarkable, except for an elevated erythrocyte sedimentation rate. Review of the March 2006 CT scan demonstrated a retroperitoneal mass encasing the aorta and involving a 3-cm segment of the right mid-ureter, resulting in moderate hydronephrosis and a delayed nephrogram. The left ureter appeared uninvolved, and there was no left hydronephrosis.
then docked to the patient and, using hook cautery and DeBakey forceps, the line of Toldt was incised from the liver to the true pelvis. The colon was then mobilized medially to the vena cava and iliac vessels to expose the retroperitoneum and right ureter. The ureter was noted to be dilated proximally, down to the level of the iliac vessels, where extensive fibrosis was encountered. Normal ureter was identified, the console surgeon, using Maryland bipolar forceps (Arrow Medical, Libertyville, IL) in the left hand and Pott scissors in the right, bluntly and sharply liberated the ureter circumferentially from the fibrotic mass. The side surgeon, using a combination of retraction on the Vesselloops and suction, provided exposure. Two specimens of the fibrotic mass were then submitted to pathology for frozen analysis. Frozen section pathological review demonstrated dense fibrotic tissue with chronic inflammation, supporting a diagnosis of retroperitoneal fibrosis. Additional specimens were sent for permanent analysis.

The robot was then undocked and removed from the field. Pure laparoscopic techniques were then applied to isolate a segment of the omentum using the LigAsure system (Valleylab, Boulder, CO). The omentum was circumferentially wrapped around the diseased portion of the right ureter. The omentum was secured using Weck clips (Pilling Weck Canada, Markham, Ontario) to the side wall, and thus the right ureter was lateralized and wrapped. A #10 Jackson-Pratt drain was placed laparoscopically adjacent to the surgical bed, a 16-French Foley catheter was left in the bladder, and the patient’s wounds were closed. He tolerated the procedure well.

The patient spent 1 night in a postoperative unit with an intravenous patient-controlled anesthesia machine, which was discontinued in the morning. Thereafter, the patient required 2 acetaminophen and oxycodone tablets 2 to 3 times daily until his discharge. The Foley catheter was removed on postoperative day (POD) 2, and the Jackson-Pratt drain was removed on POD 3, the same day the patient was discharged. The patient met discharge criteria on POD 2 but elected to remain in the hospital to be seen by the consulting rheumatologist the next day.

The patient returned for follow-up 2 weeks after surgery and described “being totally back to normal,” denying any back or flank pain or any other symptoms. He no longer required pain medication at home. Ultrasound revealed a properly positioned right ureteral stent and no hydronephrosis. The final pathologic evaluation demonstrating dense fibrotic tissue with chronic inflammation was consistent with idiopathic RPF (Figure 4). One month after surgery, the right ureteral stent was removed. Two months postoperatively, a renal scan and CT scan demonstrated resolution of the right hydronephrosis, no evidence of obstruction, and a lateralized right ureter.

Discussion

Idiopathic RPF, once thought to result from a local inflammatory reaction against antigens in the atherosclerotic plaques of the abdominal aorta, is now believed to be a manifestation of a systemic autoimmune or inflammatory disease. Recent studies also show that idiopathic RPF is significantly associated with the HLA-DRB1*03 allele, which is linked to many autoimmune diseases, such as systemic lupus erythematosus and type 1 diabetes mellitus. It is now generally accepted that the pathogenesis of this disease is multifactorial, with even the possibility of environmental risk factors, such as asbestos.

Clinical manifestations of RPF range from generalized symptoms (fatigue, fever, weight loss, and edema) to specific complaints and signs (pain and hypertension) related to the encasement or compression of adjacent structures and tissues. Because the early symptoms are nonspecific, diagnosis is usually made when the disease has progressed to cause ureteral

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obstruction or impingement of the great vessels or nerves. It is believed that symptoms before diagnosis can persist from 4 to 12 months. Laboratory studies almost always reveal an elevated erythrocyte sedimentation rate and C-reactive protein. The creatinine can be elevated from ureteral obstruction, and a mild to moderate anemia can be present secondary to chronic inflammation and renal insufficiency.

The presumptive diagnosis is usually made after imaging studies have been performed. Excretory or retrograde pyelography, ultrasonography, CT, and MRI are all commonly used in the evaluation for RPF. Currently, CT and MRI (especially in patients with elevated creatinine) are the preferred methods of imaging for this process, but neither is able to unequivocally differentiate RPF from a malignant process. CT scans depict idiopathic RPF as a homogenous plaque, isodense with muscle, which surrounds the lower abdominal aorta and iliac arteries, often encasing the ureter(s) and vena cava. MRI allows improved definition against the surrounding tissues, and RPF appears hypointense on T1-weighted images. On T2-weighted images, early/active RFP appears hyperintense, due to tissue edema and hypercellularity, whereas advanced RPF appears hypointense.

Pathologically, RPF appears grossly as a plaque-like fibrotic mass, and histologically as a nonspecific inflammatory process that varies depending on the stage of the disease. Early RPF consists of collagen bundles with capillary proliferation and abundant inflammatory cells, whereas later stages are marked by a relatively acellular and avascular fibrotic process. Unlike lymphoma, there are many different types of cells (macrophages, plasma cells, eosinophils, and lymphocytes) seen on microscopic evaluation of idiopathic RPF tissue. Only the demonstration of tumor cells within the fibrotic mass can indicate malignancy as a possible cause of RPF.

Treatment of idiopathic RPF requires halting the progression of the fibroinflammatory reaction (using steroids, immunosuppressants, tamoxifen, and other drugs) and relieving the obstruction of the ureter(s) and other retroperitoneal structures (by surgical procedures). Preventing disease recurrence or relapse requires careful patient follow-up, with regularly scheduled laboratory evaluation and imaging. Treatment of secondary RPF involves removing or treating the inciting cause of fibrosis (drug withdrawal, treatment of infection, or underlying malignancy).

Since its first description, RPF has been an insidious disease process with the potential to cause severe damage. Therapeutic intervention consists of medical and surgical therapies aimed at preserving renal function, minimizing disease progression, and possibly causing regression of the fibrotic process. Because much remains to be elucidated about the pathogenesis, natural history, and effective treatment of RPF, intense discussion persists regarding the optimal management of this disease. Current controversies in the treatment of RPF include performing deep tissue biopsies, prophylactic treatment of the uninvolved contralateral ureter, and open versus minimally invasive surgical approaches to ureterolysis.

In 1979, Lepor and Walsh reviewed the world literature on idiopathic RPF and concluded that deep tissue biopsies are required to make the diagnosis. They found it difficult to justify not performing ureterolysis, given that the success rates were as high as 90%. Moreover, they asserted that medical therapy alone is indicated only when all studies are consistent with idiopathic RPF and the operative risks are too high. By contrast, in 2006, Vaglio and colleagues found surgery to be useful only in refractory cases and argued that deep biopsies are not needed if the clinical, laboratory, and imaging findings support a diagnosis of idiopathic RPF. Because surgical treatment does not address the systemic manifestations or halt disease progression, they recommended a conservative approach with placement of nephrostomy tubes or ureteral stents, followed by medical therapy. We have found that performing numerous deep tissue biopsies is an essential component to reaching a definitive diagnosis of RPF. Once this definitive diagnosis has been made and malignancy ruled out, the medical aspects of RPF treatment can proceed without any hesitation.

By not performing prophylactic contralateral ureterolysis in any of their unilaterally diseased patients, Fugita and colleagues (in their 2002 laparoscopic ureterolysis series) challenged the notion held by many investigators that performing bilateral ureterolysis in the presence of unilateral disease is warranted. None of Fugita and colleagues’ patients with unilateral disease developed contralateral ureteral involvement at a mean follow-up of 23 months. Bilateral ureterolysis using a laparoscopic approach with or without robotic assistance requires repositioning the patient and access ports. We agree that prophylactic laparoscopic contralateral ureterolysis is not necessary and may lead to increased complications. Should a patient ever develop contralateral disease, the minimally invasive robotic approach to ureterolysis that we have described could be performed, without much morbidity.

Before 1992, open ureterolysis and ureteral transposition (with or without omental wrapping) was the standard technique to protect ureters involved in RPF. The utility of laparoscopy for
Ureteral Obstruction Due to Idiopathic RPF continued

the surgical management of this disease has been shown to avoid the morbidity associated with an open procedure. Fugita and colleagues reported an 85% success rate, with 92% of their patients remaining free of ureteral obstruction after a mean follow-up of 30 months from surgery. The mean operative time for unilateral ureterolysis was 192 minutes, mean estimated blood loss was 235 mL, and mean length of hospital stay was 4.0 days. There was 1 conversion for iliac vein injury in these unilateral patients. In 2006, Brown and colleagues described their experience with hand-assisted laparoscopic ureterolysis (HALU) as a less technically challenging and minimally invasive approach for the treatment of RPF. In their series, 5 bilateral HALU cases were performed, with a mean operative time of 259 minutes, mean estimated blood loss of 80 mL, and operative time 243 minutes, estimated blood loss 10 mL, length of hospital stay 3 days. We currently perform all of our ureterolysis procedures with the da Vinci robot and plan to report on our experience in the near future.

The treatment of RPF remains a controversial subject, with many options available to the health care provider. Although the low incidence of this disease likely precludes the possibility of any prospective randomized comparative trials, new minimally invasive treatment approaches, such as robotic ureterolysis, seem to be appealing alternatives. Our patient has been symptom free at 2 months after surgery, but longer follow-up is needed to better compare this technique with other surgical techniques.

The prognosis of idiopathic RPF is excellent, with 5-year survival rates of 90% to 100%. No predictors of response to steroid or medical therapy have been identified, and the relapse rate after discontinuation of this therapy is not established owing to the limited follow-up in most of the published studies. Ureteral obstruction is estimated to recur in 50% of patients who undergo surgery alone, and in 10% of those also treated with medical therapy. Thus, lifelong follow-up is important.

References