An Atypical Presentation of Retroperitoneal Fibrosis

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Key Words
Hydronephrosis • Idiopathic fibrosis • Infliximab • Mesenteric sclerosis • Retroperitoneal fibrosis

Abstract
A 69-year-old man with a psoriatic arthritis treated with infliximab for 1 month presented to the urology clinic for lower urinary tract symptoms. He was found to have a new diagnosis of elevated creatinine. Computed tomography of the abdomen and pelvis revealed bilateral severe hydronephrosis with abnormal soft tissue thickening of the right renal pelvis and proximal ureter. Bilateral stents were placed after ureteroscopy demonstrated no abnormalities. A computed tomography-guided biopsy of the peri-ureteral lesions revealed fibroadipose tissue with sclerosis and extensive chronic inflammation consistent with retroperitoneal fibrosis. Infliximab was discontinued and the patient was started on corticosteroids. Follow-up magnetic resonance imaging of the abdomen and pelvis at 2 months revealed total resolution of soft tissue and inflammation along the proximal ureter bilaterally. Repeat imaging demonstrated no hydronephrosis after stents removal, and the patient’s creatinine remains normal at 12 months follow-up.

Introduction
Retroperitoneal fibrosis (RPF) is part of a spectrum of idiopathic inflammatory and fibrotic processes that occurs in the retroperitoneal space, typically surrounding the infrarenal portion of the abdominal aorta, inferior vena cava, and iliac vessels. In this report, we describe an atypical imaging presentation of RPF, which involved the renal hilum bilaterally while sparing the renal, aortic, and iliac vessels. This resulted in bilateral hydronephrosis and elevated creatinine that required bilateral ureteral stent placements for decompression of the collecting system.

Case Report
A 69-year-old man with a psoriatic arthritis treated with infliximab for one month presented to the urology clinic for lower urinary tract symptoms. He was found to have a new diagnosis of elevated creatinine. Retroperitoneal ultrasound demonstrated new onset bilateral hydronephrosis. A computed tomography (CT) of the abdomen and pelvis revealed bilateral severe hydronephrosis with abnormal soft tissue thickening of the right renal pelvis and proximal ureter (fig. 1). This was also clearly visualized on magnetic resonance imaging (MRI) of the abdomen. Differential diagnosis included infection, lymphoma, amyloidosis, sarcoidosis, tuberculosis, and bilateral transitional cell carcinoma of the ureter. Bilateral flexible ureteroscopy demonstrated no abnormalities, and bilateral double J stents were placed to decompress the collecting system. Urine cytology revealed atypical cells, suspicious for carcinoma. A CT-guided biopsy of the ureteral lesions revealed fibroadipose tissue with sclerosis and extensive chronic in-
flammation concerning for an inflammatory and fibrotic process. Immunostain for IgG4 antibody revealed occasional plasma cells, but no increase in plasma cell percentage. Upon further review of the patient’s medical history, he was started on infliximab for psoriatic arthritis a month prior to the new onset bilateral hydronephrosis (the patient previously had a normal retroperitoneal ultrasound). Infliximab was discontinued, and the patient was started on corticosteroids. Follow-up MRI of the abdomen and pelvis in 2 months revealed total resolution of the inflammation along the proximal ureter bilaterally (fig. 2). Repeated retroperitoneal ultrasound following stents removal revealed no hydronephrosis and the patient’s creatinine remains normal at 24 months follow-up.

Discussion

RPF was first described by Albarran in 1905, and subsequently reported in the English literature by Ormond in 1948. Idiopathic RPF, also coined Ormond’s disease accounts for two-thirds of cases reported. Secondary causes of RPF include drugs (derivatives of ergot alkaloids), neoplasms (lymphoma, metastatic disease, retroperitoneal sarcoma, carcinoïd tumor, primary cancers of the stomach, colon, lung, breast, genitourinary tract or thyroid gland), infection, post radiation therapy, trauma, history of abdominal surgeries and retroperitoneal hemorrhage/hematoma [1, 2]. The underlying pathophysiology remains unknown as only a paucity of case reports and small case series are available in the literature. RPF is not histologically well defined and it can be extremely challenging to differentiate RPF from other fibrous tissue-producing disorders such as sclerosing mesenteritis, isolated periaortitis, and inflammatory abdominal aortic aneurysm.

The clinical features for RPF are nonspecific, making it a diagnosis of exclusion. Clinical manifestations include lower abdominal or flank pain, weight loss and constipation. Patients may also experience nausea and/or vomiting, altered consciousness due to anuria from renal failure. The involvement of renal vessels can also result in renovascular hypertension. Compression of retroperitoneal lymphatic vessels and veins can also result in edema of the lower extremities, scrotal swelling and varicoceles [3].

Due to its protean of symptoms, laboratory studies are typically nonspecific. Inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein level, may be elevated. Radiological investigative studies are keys in establishing a diagnosis. CT and MRI of the abdomen and pelvis remains the most sensitive imaging modality for diagnosing RPF. RPF typically involves the aorta and ranges from the origin of the renal arteries up to the bifurcation of the common iliac arteries [4].

In our patient, the presentation of RPF was atypical as seen on CT of the abdomen and pelvis. Soft tissue masses were seen around the renal pelvis bilaterally concerning for lymphoma and urothelial cell carcinoma of the renal collecting system. Ureteroscopy was performed to rule out primary urothelial cell carcinoma of the ureter. Following a negative ureteroscopy, a biopsy of the mass was warranted to obtain tissue diagnosis. This confirmed the diagnosis of an ongoing inflammatory process. RPF and mesenteric sclerosis is indistinguishable under microscopy. However, mesenteric sclerosis typically do not present around the renal pelvis. Hence, this patient had an abnormal presentation of what is believed to be RPF.

To date, there have been 2 cases of infliximab reported which may have resulted in RPF. Both patients were above the age of 60, similar to our patient. Ironically, in-

Fig. 1. CT of the abdomen and pelvis (contrast phase). Arrow depicts chronic inflammation and fibroadipose tissue.
Retroperitoneal fibrosis is an investigative therapy for refractory RPF, which has demonstrated success in a patient who had idiopathic RPF that was refractory to steroids [5]. RPF remains a rare disease that can affect a patient anywhere in the retroperitoneal space. Clinicians should include RPF as part of their differential diagnosis to ensure that a prompt diagnosis can be established and appropriate treatment provided to prevent avoidable consequences.

**References**

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