CLINICAL VIGNETTE

A Rare Cause of Bilateral Ureteral Obstruction

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Case Report

A 48-year-old female presented to the emergency room with several weeks of generalized abdominal discomfort that progressed over the last 5 days. Her abdominal discomfort began as a sensation of constipation with new onset gastroesophageal reflux for which she tried over the counter antacids and laxatives. Five days prior to presentation, her abdominal pain increased in intensity and described as a dull pressure in the upper abdominal region with radiation to the back. The pain was relieved when she curled into a fetal position and was not relieved with ibuprofen. She was not taking any other medications. She also reported an intentional 25 lbs weight loss over the past 6 months while dieting. She denied nausea, vomiting, or changes in bowel habits.

Her physical exam was notable for significant tenderness over her epigastrium and bilateral flank areas. Her complete blood count had no significant abnormalities. A complete metabolic panel revealed a normal blood urea nitrogen (BUN) of 18 mg/dL and creatinine of 0.92 mg/dL. A urinalysis was unremarkable. Computed tomography (CT) of her abdomen and pelvis revealed bilateral hydronephrosis with extensive retroperitoneal and peritoneal inflammatory changes tracking along the retroperitoneal and mesenteric vasculature (Figure 1). She was subsequently admitted for bilateral ureteral stent placement after retrograde pyelogram and cystoscopy by Urology. An evaluation for secondary causes of her retroperitoneal fibrosis was started prior to discharge. Malignancy was of low suspicion based her imaging. Other laboratory results were notable for an elevated ANA titer of 1:160. Her C-reactive protein and estimated sedimentation rate (ESR) were also elevated at 42.2 mg/dL and 44mm/hr respectively.IgG subset was within normal limits. A core biopsy of the lesion revealed interwoven and anastomosing bands of hyalinized fibrous tissue with inflammatory cell infiltrate suggestive of retroperitoneal fibrosis. No significant IgG4 positive plasma cells were noted on biopsy. Following comprehensive oncologic and rheumatologic outpatient evaluation to rule out secondary causes of fibrosis, she was started on a prednisone taper and mycophenolate mofetil 1.5 g twice per day for idiopathic retroperitoneal fibrosis.

Discussion

Idiopathic retroperitoneal fibrosis is a rare disorder characterized by fibroinflammatory tissue involving retroperitoneal structures including the ureters, abdominal aorta, and inferior vena cava. Onset is often insidious and is only detected after significant fibrotic invasion. The most common clinical manifestation is lower back, flank, or abdominal pain secondary to ureteral encasement. The disease has an estimated incidence of 0.1-1.3 cases per 100,000 persons per year and a prevalence of 1.4 cases per 100,000 inhabitants.[1] Because of its rarity combined with its vague presentation, the fibrosis is often extensive at the time of diagnosis with associated hydronephrosis, impaired renal function, and renal atrophy.[1-3] Additional urologic manifestations include chronic pelvic and testicular pain as well as varicoceles and hydroceles due to testicular vein encasement. Chronic inflammation of the aorta, iliac arteries, and inferior vena cava may cause lower extremity claudication and edema at the time of presentation.

The differential diagnosis for the etiology of retroperitoneal fibrosis includes a wide array of medications including methyldopa, ergot alkaloids, beta blockers, hydralazine, and infliximab. Neoplastic causes include metastatic disease, lymphomas, and sarcomas. Infectious causes include tuberculosis, histoplasmosis, actinomycosis, and syphilis. Fibrotic changes can also be secondary to postoperative or post-radiation changes.[4] However, idiopathic retroperitoneal fibrosis represents up to two-thirds of diagnoses where no primary cause is identified. The disease state is associated with other autoimmune conditions, the most common being IgG4-related disease which is characterized by a dense lymphoplasmacytic infiltrate with a high percentage of $IgG4^+$ plasma cells. The infiltrate can lead to multisystem fibrosis, with a constellation of resultant conditions including autoimmune pancreatitis, Riedel's fibrosing thyroiditis, and noninfectious aortitis.[5]

The goal of treatment is to relieve obstruction of retroperitoneal structures and to slow further fibrotic inflammatory changes in the retroperitoneum. Management of idiopathic retroperitoneal fibrosis can be divided into acute and chronic therapies. Acutely, the patient should be evaluated for ureteral obstruction and vascular inflammation with ultrasonography, CT or MRI. Subsequent ureteral and endovascular stenting should be performed if appropriate. Additionally, symptomatic patients should be evaluated for deep vein thrombosis and pulmonary embolism given the condition's potential for venous encasement. Laboratory studies to investigate for infections, malignancies, and further autoimmune diseases such as rheumatoid disease and vasculitides are indicated along with biopsy of the retroperitoneal mass to confirm diagnosis. Thereafter,

disease control is achieved using steroid tapers and other immunsuppressants including mycophenolate mofetil, azathioprine, and cyclophosphamide. Tamoxifen, a selective estrogen-receptor modulator, has also been shown to have anti-fibrotic activity, though it is not as effective as steroids in achieving prevention of relapses.[6] Additionally, periodic imaging of the thoracic and abdominal aorta as well as the iliac arteries should be performed to assess for aneurysmal degeneration and possible need for repair.

Conclusion

Idiopathic retroperitoneal fibrosis presents a diagnostic challenge because of its rarity and nonspecific symptoms. Awareness of retroperitoneal anatomy in relation to presenting symptoms aids in detection of the disease, with imaging remaining a major asset in the diagnosis.

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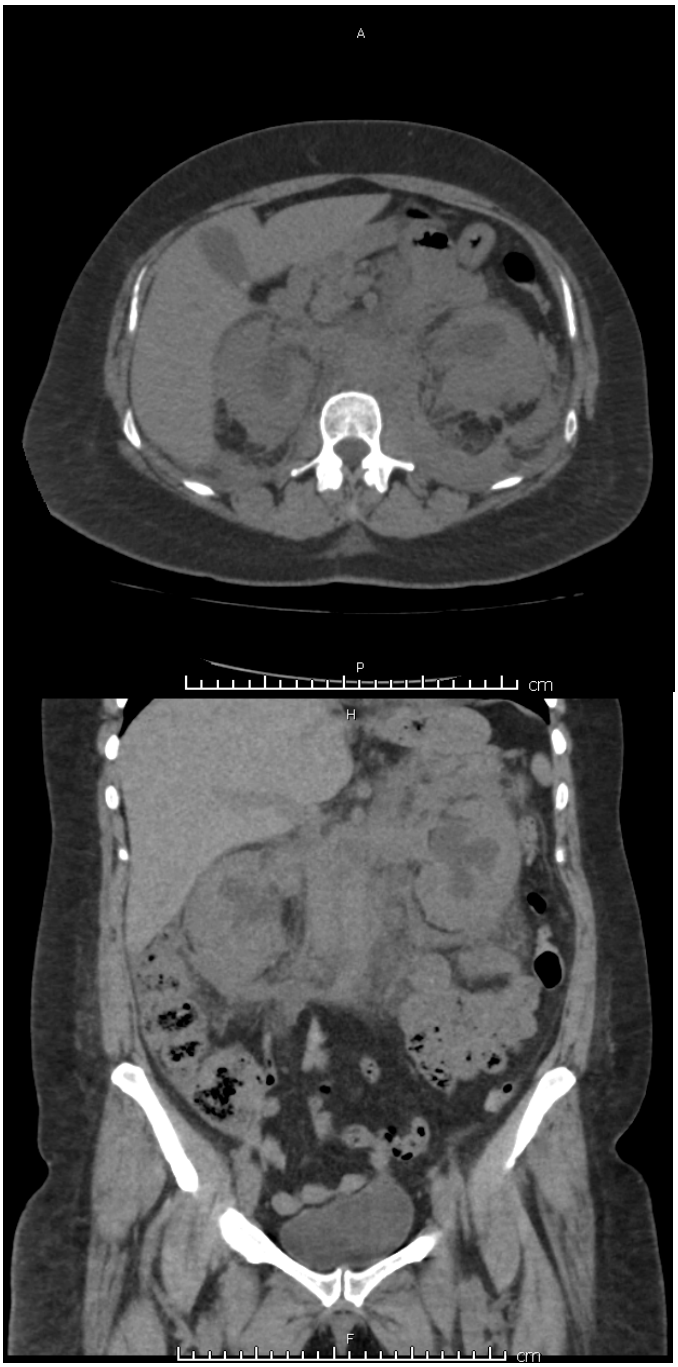

Figure 1. CT abdomen and pelvis without contrast axial and coronal views demonstrating bilateral hydronephrosis and confluent mass-like opacities encasing the aorta, inferior vena cava, mesenteric vessels, and proximal ureters.