Images

Retroperitoneal fibrosis

Michael A. Bellamy, DO*; Eric B. Tomich, DO*

A 57-year-old man presented to the emergency department with a chief complaint of progressive left lower quadrant abdominal and lower back pain for the past 3 weeks. Review of systems revealed only a history of constipation. There was moderate tenderness to palpation of the left abdomen on examination. Vital signs were normal. A complete blood count, basic metabolic panel, and liver function tests with lipase were normal except for platelets of 479,000/mm^3 and creatinine of 1.2 mg/dL (glomerular filtration rate 74 mL/min). Computed tomography (CT) of the abdomen and pelvis with intravenous contrast (Figure 1) was performed, followed by bedside ultrasonography (US) of the aorta (Figure 2). The bedside US was performed to gain images of possible abnormal findings seen on aortic US. After identification of a mass surrounding the aorta and discussion with Vascular Surgery, the erythrocyte sedimentation rate and C-reactive protein were obtained, with values of 54 mm/hr and 6.2 mg/dL, respectively. The final diagnosis was based on imaging studies including magnetic resonance imaging (MRI). A biopsy of the tissue was not attempted due to the risk associated with biopsy. The patient was placed on high-dose prednisone (1 mg/kg/day), which resulted in the retroperitoneal fibrosis (RPF) shrinking by one-half after a month of treatment.

RPF is a rare condition resulting in chronic inflammation and fibrosis of the tissue surrounding the aorta and adjacent retroperitoneal structures.[1,2] Two-thirds of cases are idiopathic but hypothesized to be either a manifestation of systemic autoimmune disease or an exaggerated local inflammatory response to aortic atherosclerosis. The autoimmune diseases are generally connected to the HLA-DRB1*03 allele, which is associated with type 1 diabetes mellitus, myasthenia gravis, and systemic lupus erythematosus.[2,3] The remaining cases are secondary to malignancy, infections, trauma, radiotherapy, surgery, or medications. The most common medications implicated are ergot alkaloids such as methysergide, beta-blockers, and dopamine agonists.[1,2,4] The incidence and prevalence of secondary causes are unknown, other than malignancy, which is the cause in approximately 8% of cases.[2,4,5]

The presentation is vague and most commonly consists of progressive pain in the lower back in approximately 38%, flank in 34%, and abdomen in 23 to 40% of patients. Less common symptoms include weight loss and scrotal pain. Oliguria may herald ureteral obstruction, whereas vascular compromise may manifest as lower extremity edema, claudication symptoms, or intestinal ischemia.[1,5] Inflammatory markers such as erythrocyte sedimentation rate and C-reactive protein are elevated in 80 to 100% of cases.[1,6] Renal function studies are important if there is ureteral involvement.

CT and MRI are the most sensitive diagnostic modalities and help rule out secondary etiologies and aortic wall involvement. No studies have identified which of the two is superior. CT usually reveals a well-demarcated, irregularly shaped mass in the retroperitoneum that is isodense to muscle with noncontrast studies. The degree of enhancement with intravenous contrast studies correlates with the inflammatory activity of the process. Typically, the greatest fibrosis is centered at the aortic bifurcation with involvement of the iliac vessels. In contrast to malignancies, such as lymphoma, bone destruction and displacement of the abdominal aorta and vena cava anteriorly from the

From the *Department of Emergency Medicine, San Antonio Military Medical Center, Fort Sam Houston, TX.

Correspondence to: Dr. Michael A. Bellamy, Department of Emergency Medicine, San Antonio Military Medical Center, Fort Sam Houston, TX 78234; mbellamydo@gmail.com.

This article has been peer reviewed.

 CJEM 2014;16(1):63-64 DOI 10.2310/8000.2013.131009

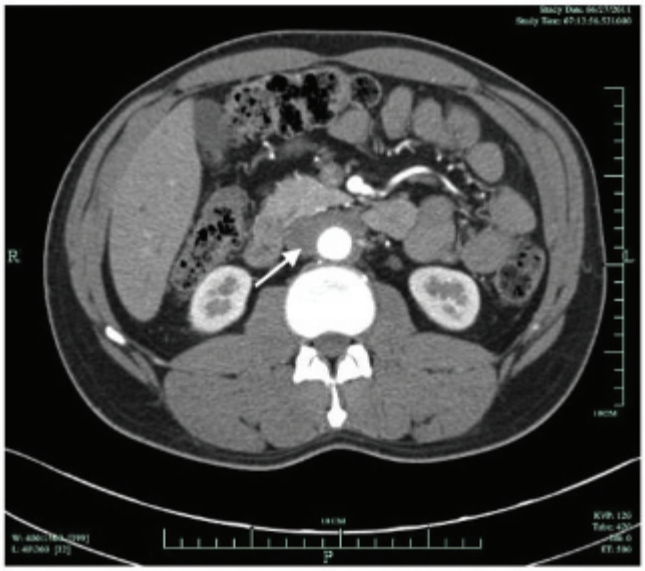

Figure 1. Computed tomographic scan of the abdomen and pelvis with intravenous contrast. The *arrow* points to an irregularly shaped mass adjacent to the aorta.

spine are rare in RPF. US will demonstrate a hypoechoic, periaortic mass with possible hydronephrosis. US can be used to monitor the disease. MRI demonstrates characteristics similar to those of other fibrotic processes. T_1-weighted imaging produces low signal intensity, whereas T_2-weighted signal of this tissue varies with the degree of active inflammation.[2,3]

RPF should be considered in the differential diagnosis for patients with vague abdominal and back complaints as delayed diagnosis may result in greater morbidity. Significant renal or vascular involvement can require inpatient hospitalization and surgical management. Corticosteroids have been shown to improve symptoms and lead to a reduction in the size of the mass and resolution of obstructive complications. Immunosuppressive agents have proven beneficial in patients with contraindications or poor clinical results with glucocorticoids.[1,2,7,8]

Competing interests: None declared.

Keywords: abdominal, aortitis, fibrosis, imaging, retroperitoneal

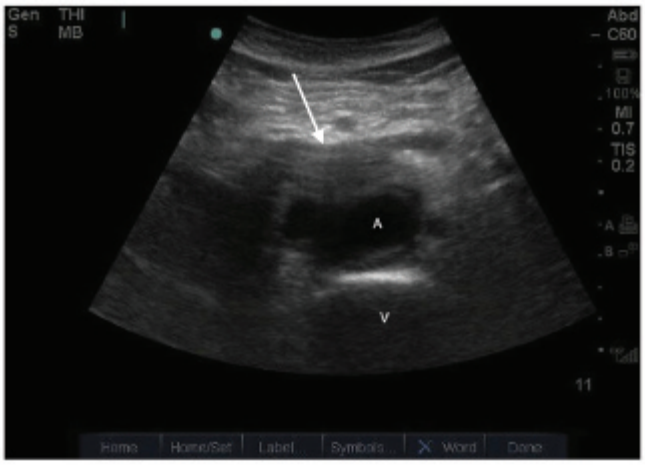

Figure 2. Bedside sonogram of the aorta. The *arrow* points to the border of a hypoechoic, periaortic mass. A = aorta; V = vertebral body.

REFERENCES

1. Vaglio A, Salvarani C, Buzio C. Retroperitoneal fibrosis. *Lancet* 2006;367:241-51, doi:10.1016/S0140-6736(06)68035-5.
2. Vaglio A, Palmisano A, Corradi D, et al. Retroperitoneal fibrosis: evolving concepts. *Rheum Dis Clin North Am* 2007;33:803-17, doi:10.1016/j.rdc.2007.07.013.
3. Martorana D, Vaglio A, Greco P, et al. Chronic periaortitis and HLA-DRB1*03: another clue to an autoimmune origin. *Arthritis Rheum* 2006;55:126-30, doi:10.1002/art.21698.
4. Cronin CG, Lohan DG, Blake MA, et al. Retroperitoneal fibrosis: a review of clinical features and imaging findings. *AJR Am J Roentgenol* 2008;191:423-31, doi:10.2214/AJR.07.3629.
5. Koep L, Zuidema GD. The clinical significance of retroperitoneal fibrosis. *Surgery* 1977;81:250-7.
6. Vaglio A, Corradi D, Manenti L, et al. Evidence of autoimmunity in chronic periaortitis: a prospective study. *Am J Med* 2003;114:454-62, doi:10.1016/S0002-9343(03)00056-1.
7. van Bommel EF, Siemes C, Hak LE, et al. Long-term renal and patient outcome in idiopathic retroperitoneal fibrosis treated with prednisone. *Am J Kidney Dis* 2007;49:615-25, doi:10.1053/j.ajkd.2007.02.268.
8. Kardar AH, Kattan S, Lindstedt E, et al. Steroid therapy for idiopathic retroperitoneal fibrosis: dose and duration. *J Urol* 2002;168:550-5, doi:10.1016/S0022-5347(05)64677-0.