

Spontaneous resolution of idiopathic aortitis and pitfalls in diagnosis

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ABSTRACT

Idiopathic aortitis is an inflammatory disease of the aorta that is diagnosed after the less frequent infectious and rheumatologic variants are excluded. The etiology and natural history of the disease are poorly understood, and its presentation is variable; the need for exclusion of infectious, malignant, and rheumatologic causes can make its evaluation and diagnosis challenging. Treatment is tailored to the diagnosis and may include observation, antimicrobial therapies, and immunosuppressive agents when appropriate. Operative therapy is rarely needed and reserved for symptomatic patients or instances in which infection cannot be excluded. We present a case of idiopathic aortitis that resolved spontaneously with expectant management and discuss the pitfalls in the diagnosis and care of the disease. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:95-8.)

Keywords: Aortitis; Large vessel vasculitis; Nonaneurysmal; Noninfectious; Idiopathic

Aortitis represents a diverse set of pathologic processes characterized by inflammatory changes to the aortic wall or the periaortic tissue. The disease is categorized into infectious, rheumatologic (including autoimmune), and idiopathic variants, which may have similarities in symptoms and imaging characteristics but can usually be separated with careful use of serum markers and blood cultures. Of the pathologic specimens from patients undergoing aortic operations, the most prevalent type of aortitis is idiopathic, representing approximately 70% of the cases with evidence of aortic inflammation.^{1,2} Early diagnosis of infectious aortitis is critical as untreated infection can result in significant complications, such as aortic rupture and sepsis.³ We present a case of idiopathic aortitis (IA) that required a prolonged diagnostic evaluation and spontaneously resolved with expectant management. Informed consent for publication was obtained from the patient.

CASE REPORT

A 62-year-old man with history of polysubstance abuse, hypertension, hypercholesterolemia, and stage 3 chronic kidney disease presented to his primary physician in

July 2016 secondary to abdominal pain. The patient was treated for presumed urinary tract infection; however, his urine culture was eventually found to be negative. The patient was then treated with a bowel regimen for possible constipation; however, the symptoms failed to improve, and he presented to his primary physician again 7 weeks later. A non-contrast-enhanced computed tomography (CT) scan of abdomen and pelvis (Fig 1) obtained in August 2016 was read to have findings of a suspected infrarenal abdominal aortic aneurysm measuring 4.1 cm with "worrisome features for dissection." The patient underwent subsequent CT angiography (Fig 2) in September 2016 that demonstrated periaortic soft tissue thickening without dissection or aneurysmal changes and with the presence of left-sided hydronephrosis. The urology service was consulted, and a ureteral stent was placed approximately 2 months after the initial presentation. CT-guided biopsy was attempted a month later to obtain a tissue specimen because of the concern of underlying malignant disease, but the procedure was discontinued secondary to the inability to access the lesion safely.

The patient's symptoms of abdominal discomfort had largely subsided after placement of the ureteral stent, but because of the persistent periaortic mass 6 months later, the patient was referred to the general surgery service. His case was discussed at tumor board. Serum tumor markers, scrotal ultrasound, and magnetic resonance imaging (MRI) were recommended to further evaluate the periaortic mass; the differential diagnosis consisted of germ cell tumor, retroperitoneal fibrosis, lymphoma, sarcoma, and neuroendocrine tumor. Serum tumor markers and scrotal ultrasound failed to elucidate the cause of the mass, and the patient was referred to the vascular surgery clinic a month later in May 2017 for evaluation of questionable claudication. The leg pain was determined to be secondary to sciatica. Further examination of the patient revealed normal findings on abdominal examination and no complaints of abdominal symptoms, but a review of his previous CT imaging raised concern for aortitis. A repeated CT scan was obtained and the rheumatology service was consulted. The radiologist

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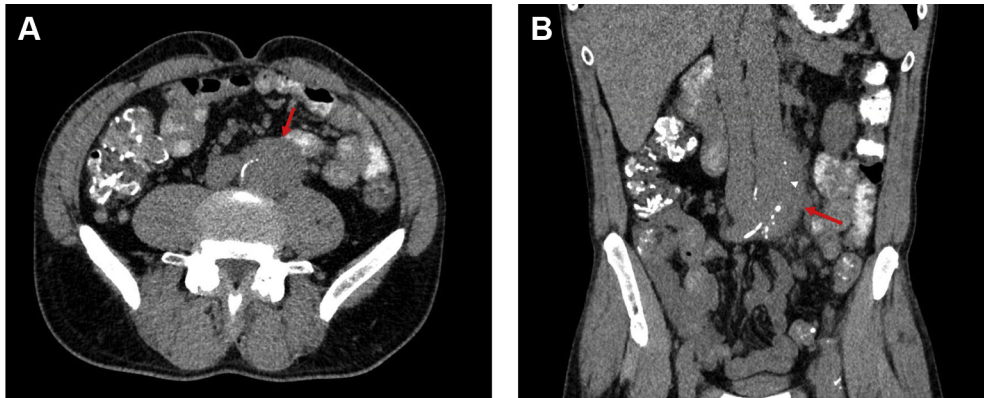


Fig 1. Axial (A) and coronal (B) views of initial abdominopelvic computed tomography (CT) performed for workup of abdominal pain. The arrows point to what was believed to be an aneurysmal dilation of the infrarenal abdominal aorta.

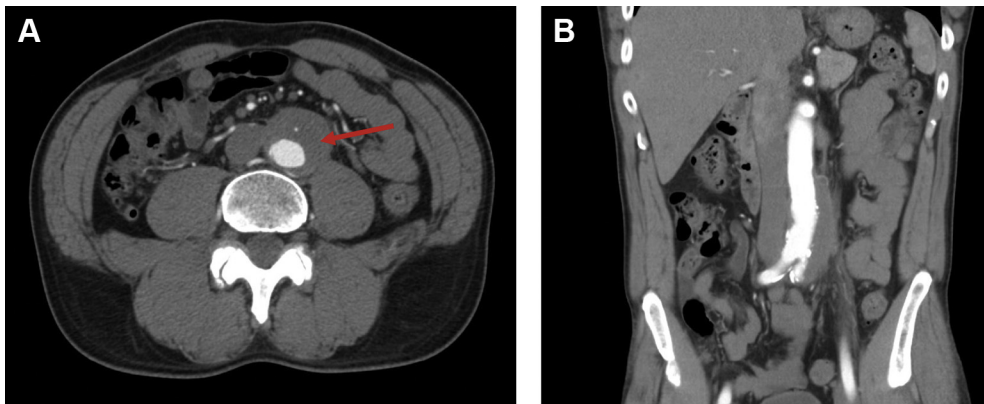


Fig 2. Axial (A) and coronal (B) views of abdominopelvic computed tomography (CT) angiography to evaluate the abdominal aortic aneurysm. A significant periaortic thickening is seen in the anterolateral area (arrow).

interpreted the findings of the new CT scan as “periaortic soft tissue as was previously seen” and noted that the diameter of the periaortic soft tissue had decreased to 3.2 cm from the measurement of 4.2 cm seen on the CT scans of September 2016. The rheumatology team was in agreement of obtaining the previously suggested MRI and added cultures and expanded the evaluation to rule out infectious, malignant, and autoimmune causes. Special laboratory evaluations including C-reactive protein, total protein, serum protein electrophoresis, hepatitis panel, proteinase 3 antibodies, myeloperoxidase antibody, immunoglobulin G (IgG) and IgG subclasses panel (including IgG4), anti-thyroid peroxidase, perinuclear and cytoplasmic antineutrophil cytoplasmic antibodies, quantiferon, histoplasma antigen, and human immunodeficiency virus screen were nonrevealing. MRI confirmed periaortic thickening measuring approximately 5 mm (Fig 3) that was consistent with aortitis. No aortic aneurysm or dissection was noted. Because the patient had no complaints of abdominal discomfort and a decrease in the diameter of the aortic disease, the vascular and rheumatology teams elected to observe the patient with serial imaging without immunosuppressive or antibiotic therapy. The patient was monitored with repeated CT angiography in November 2017 (Fig 4) that demonstrated a resolution of his periaortic thickening. With

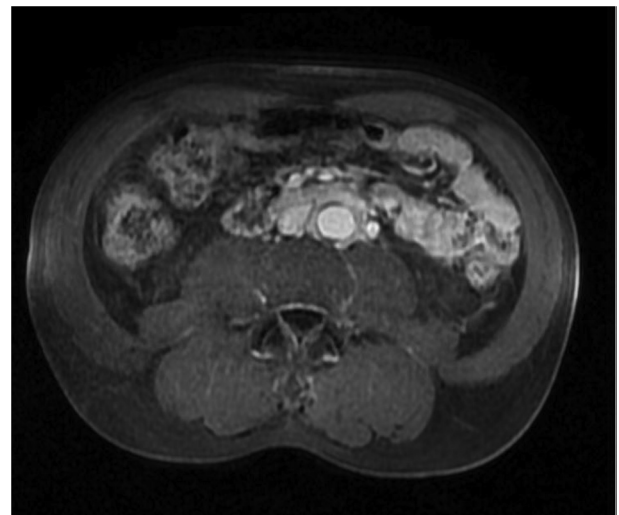


Fig 3. Magnetic resonance imaging (MRI) of the abdomen with circumferential periaortic thickening suggestive of aortitis.

resolution of the inflammation, the left ureteral stent was removed. The patient was seen again 6 months later without further symptoms and without recurrence of hydronephrosis.

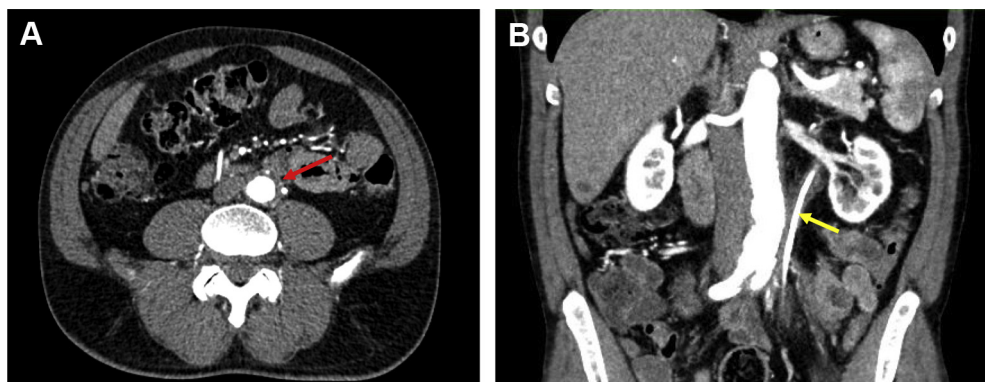


Fig 4. Axial (A) and coronal (B) views of abdominopelvic computed tomography (CT) angiography showing resolution of periaortic inflammation approximately 1 year after the initial diagnosis. The red arrow points to the resolution of periaortic thickening. The yellow arrow points to the ureteral stent.

DISCUSSION

The prevalence of aortitis has been reported to be around 4% to 6% in a pathologic analysis of aortic specimens from two surgical cohorts.^{1,2} Risk factors for the development of IA include a history of connective tissue disease, advanced age, and diabetes.¹ The pathophysiologic trigger behind the inflammatory process of IA is largely unknown. It has been shown in giant cell arteritis that the activation of adventitial dendritic cells by pathogen-derived macromolecules results in vasculitis,⁴ and such findings provide concepts that can be extrapolated to the pathophysiologic mechanism of aortitis.

The workup of a patient with suspected IA is largely focused on excluding a malignant or infectious etiology³ and systemic rheumatologic, inflammatory, and autoimmune causes, such as Takayasu arteritis, giant cell arteritis, and IgG4 periaortitis.^{5,6} Imaging with CT or MRI with intravenous administration of contrast material is critical in making the diagnosis.⁷ Contrast-enhanced imaging helps differentiate between aneurysmal diseases and periaortic soft tissue swelling as exemplified by our case. Workup should also include blood cultures and biochemical evaluation because of the consideration of possible infectious as well as rheumatologic causes.^{8,9} When possible, biopsy can be used to confirm a diagnosis, but as in this case, it is often not practical or possible.

Treatment of aortitis is usually nonoperative, with operative management reserved for symptomatic aneurysmal disease or when infection cannot be excluded. Mainstay medical treatment of IA can involve the use of steroids and immunosuppression.^{2,10} Because of the rarity of IA, optimal treatment protocols have not been established, and the natural history is poorly understood. Our patient had relatively mild symptoms and reached remission simply with time. This type of expectant management spares the patient from potential adverse effects of operative and nonoperative therapies. In our practice, we evaluate and manage patients with IA in collaboration with an expert rheumatologist. Based on

our experience, initial management consisting of close follow-up with repeated cross-sectional imaging and no immunosuppressants may be adopted in patients with no symptoms or mild and tolerable abdominal and back symptoms, normal inflammatory and autoimmune parameters, and negative findings on infectious evaluation or when the diagnosis is still questioned, especially if a biopsy is not an option. There are data supporting the benefits of the use of glucocorticoids and immunosuppression for patients with large-vessel vasculitis.¹¹ In those patients, we start with glucocorticoids either by mouth (initial agent of choice is oral prednisone) or intravenously. If steroid-sparing medications are needed, then we consider the use of medications like cyclophosphamide, azathioprine, mycophenolate mofetil, and methotrexate; there are reports that show anti-tumor necrosis factor α drugs effective in patients with refractory aortitis.¹² Retroperitoneal fibrosis with ureteral obstruction should be managed in collaboration with urology and rheumatology consultants and usually includes glucocorticoid therapy along with urologic intervention to relieve the obstruction. Patients with IA should be closely monitored for the development of complications, such as aortic aneurysm or occlusive disease. Endovascular techniques have supplanted open surgical reconstruction, and any elective surgical intervention should be deferred until attainment of disease remission for better long-term outcomes.^{13,14}

The diagnosis of IA we reached in this case may be questioned. As is often the situation, short of a tissue diagnosis, the clinical diagnosis of vasculitis is never completely certain, and the literature is replete with instances of mimics of vasculitis to include certain viruses and illicit drug-induced processes.¹⁵ In addition, the “natural history” of untreated incidental vasculitis or aortitis is not well understood as most patients are treated, and thus the resolution is attributed to the treatment regimen and not the natural history. Finally, some would argue that resolution of aortitis without treatment, as

noted in our patient, should be attributed to a diagnosis other than vasculitis; nevertheless, an alternative differential diagnosis could not be offered after consultation with various subspecialty teams. Based on our experience and the findings from previous pathologic studies,^{1,2} we suspect that the phenomenon of incidental aortitis is experienced not uncommonly, and we hope to provide a guide for cases in which treatment may not be necessary.

CONCLUSIONS

This case demonstrates the challenges of the evaluation and management of IA. As in our patient, some cases may resolve spontaneously with expectant management, obviating the potential adverse effects associated with the use of medications. Multi-institutional consortiums may help further define disease evaluation and treatment strategies in the future.

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