Aortic Root Aneurysms in Twenty-Year-Olds: Don’t Forget to Check the Histology

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Abstract

When confronted with an aortic root aneurysm in a young patient, one of the most common etiologies is a connective tissue disorder (CTD, such as Marfan’s, Loeys Dietz, and Ehlers-Danlos.) Aortitis is considered rare among young patients and can be easily missed in asymptomatic cases. Aortitis can be difficult to diagnose but is important to investigate during surgery because missing the diagnosis may affect late outcomes. Undertreated aortitis may progress beyond the treated segment. Our understanding of arteritis is improving, and it has been suggested that Giant Cell Arteritis (GCA) and Takayasu Arteritis (TAK) may represent an overlapping spectrum of disease. CTD, GCA, and TAK require lifelong surveillance and immunosuppressive treatment for active disease. However, Autoimmune Aortitis may be cured surgically. We describe two patients in their twenties with aortic root aneurysms who underwent valve sparing aortic root replacement. Both were found on histologic examination to have chronic aortitis. The cases presented here demonstrate the importance for surgeons to send aortic specimens for histologic review when performing aortic aneurysm repair.

Keywords: Aorta; Aneurysm (Root); Histology (Aortic); Pathology (Aortic); Surgery

Case Presentation

Case 1

A 27-year-old woman with a history of chronic back pain presented with an exacerbation. Computed tomography (CT) revealed a 4.5 cm ascending aortic aneurysm. Follow-up serial imaging showed diffuse dilation from the aortic root to the descending aorta with a 5.1 cm maximum diameter (Figure 1). Echocardiogram confirmed a tricuspid aortic valve with moderate-to-severe regurgitation. Family history was significant for a cerebral aneurysm in a cousin. There were no clinical signs suggestive of a CTD or vasculitis; and blood work revealed normal levels of inflammatory markers. She underwent a valve-sparing aortic root replacement with valve reimplantation (modified David’s procedure); ascending aorta and aortic arch repair as well as a prophylactic first stage elephant trunk procedure. Operative findings included a chronic, focal proximal ascending aortic dissection with interval healing. Aortic tissue histopathologic examination showed chronic aortitis (Figure 2A and 2B). She was discharged home on post-operative day 10. Rheumatologic workup found no evidence of active arteritis in other vessels that necessitated treatment. Two years later; she underwent successful elephant trunk completion with a thoracic endovascular aortic repair.

Case 2

A 23-year-old man presented with an incidentally found ascending aortic aneurysm during workup for a new heart murmur. Echocardiogram revealed a dilated aortic root and ascending aorta; and tricuspid aortic valve with trivial regurgitation. CT confirmed a proximal aortic aneurysm with 5.1 cm maximum diameter. There was no family history of aneurysms. He underwent a modified David’s reimplantation procedure with an ascending aorta and hemi-arch repair. Operative findings were again significant for a healed chronic aortic root dissection. Histopathologic review of the aorta also noted a chronic aortitis (Figure 2C and 2D). The patient was discharged home on post operative day seven. Rheumatologic workup found no evidence of other vessel involvement and no indication
for immunosuppressive therapy. He was doing well at his one year follow-up. The MRA showed a stable; well-healed aortic repair with no findings of arthritis; and a well functioning aortic valve with trivial regurgitation.

**Discussion**

Patients with asymptomatic aortitis and associated aneurysm are diagnosed after surgery based on histopathologic review demonstrating giant cells and other features of vasculitis [1]. These two cases demonstrate that it is imperative to send aortic tissue for histologic evaluation after aortic aneurysm repair; particularly for asymptomatic patients [2,3]. Aortitis may be grossly underestimated in younger patients because specimens are not routinely reviewed. In a study involving 513 patients aged 16-85 years undergoing an ascending aortic aneurysm repair; 57 were diagnosed with aortitis after histologic review [4].

Common etiologies of aortitis include GCA; TAK; and Autoimmune Aortitis. However; other important causes include infection; primary vasculitides such as Cogan’s syndrome and Behcet’s disease; as well as inflammatory diseases with spondyloarthropathies; rheumatoid arthritis; and sarcoidosis. Both of these cases show large vessel arteritis with laminar necrosis surrounded by lymphohistiocytic infiltrates and multinucleated giant cells in the media. These features are typically described in GCA which usually affects a much older population but are occasionally seen in younger patients with isolated vasculitis. The cases differ in their extent of aortic involvement; and their operations were tailored to these preoperative imaging findings.

Histopathologic features alone are not specific to predict isolated versus systemic vasculitis. While it confirms a large vessel vasculitis diagnosis; further classification requires clinical correlation and radiologic investigations. A histologic diagnosis of chronic aortitis with giant cells encompasses GCA; TAK; and Autoimmune Aortitis. GCA has a propensity to affect external carotid arteries. It can present early with symptoms; but late and more serious complications; including aneurysms and dissections; are often detected incidentally during imaging for other reasons or during autopsy. Aortic involvement has been found in up to 18% of asymptomatic cases [3]. It is not surprising that both cases described here had evidence of focal dissection found incidentally during surgery. Patients with GCA are 17.3 times more likely to develop a thoracic aortic aneurysm and 2.4 times more likely to have an isolated abdominal aortic aneurysm than normal controls [4].

It has been speculated that TAK and GCA are not two distinct diseases but represent part of a spectrum within a single entity [1] Compared to GCA; TAK is known to have a younger age of onset; a potential predilection for Asian women and aortic involvement. One study questioned these clinical distinctions and found that patients with GCA and TAK do have similar signs; symptoms; and imaging abnormalities suggesting that the difference in diagnosis may be due to bias [1].

Autoimmune Aortitis; on the other hand; is a disease that is localized to the thoracic aorta; and may not require medical therapy after surgery [5]. The mean age of onset in our early series was 63 years [2]. These patients are unique because their disease can be surgically cured before the histologic diagnosis is made; as was the case for our second patient. In an early series; we found that these patients usually did not require any further intervention; but most patients were older and late follow-up was limited [2]. In our more recent experience; we have found that many of these patients do require later thoracic aortic operations and immunosuppressive therapy does little to slow this progression [5].

Aortitis was also commonly associated with aortic valve disease in our recent series; but the etiology of valve dysfunction was due to mixed morphologic findings. The aortic cusp tissue was healthy appearing in both of these patients despite root dilatation so the intraoperative decision in these young patients was to spare the valve by modified reimplantation. Little is known about how this disease process affects the aortic valve. Time will tell whether the surgery proves to be definitive treatment in both of these patients. Given their young age and the limitation of our knowledge about the etiology of thoracic aneurysms; surveillance MRA of the thoracic aorta at least every five years was recommended.

The post-operative Histopathologic diagnosis of aortitis should be followed by comprehensive rheumatologic evaluation to rule out other vessel involvement and assess for symptoms or signs of an underlying vasculitis or CTD. These patients may benefit from additional medical treatment including corticosteroids or other immunosuppressive to prevent new aneurysms or further progression of vasculitis. Our cases demonstrate the vital importance of histopathologic assessment of surgically respected thoracic aortic tissue to facilitate comprehensive management of aortitis.
References


