Isolated Necrotizing Aortitis Presenting as Incidental Thoracoabdominal Aortic Aneurysm: A Case Report

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ABSTRACT

Noninfectious aortitis is frequently asymptomatic yet often leads to the development of ascending aortic aneurysms requiring repair. We present the case of a 64-year-old Caucasian woman who presented to our medical center with an incidentally discovered thoracoabdominal aneurysm. She had previously been in good health and had not complained of chest pain or been otherwise symptomatic. At presentation to our clinic, her ascending aorta measured 5 cm at the sinotubular junction (STJ) with dilation of her descending thoracic aorta to 6 cm. Her coronary angiogram was normal. Cross-sectional imaging was notable for thickening of the aortic wall along its length. The entity of isolated noninfectious aortitis is increasingly being recognized as an identifiable factor leading to ascending aneurysmal disease. In reviewing this case, we outline current understanding and guidelines for management and follow-up of this pathology. Operative repair of the ascending arch was conducted using an interpositional graft. The aortic valve apparatus was normal and the proximal anastomosis was created at the STJ. Intraoperatively, an inflammatory obliteration of the aortopulmonary window was noted. It was not possible to completely excise the posterior aortic wall. Our patient had an uneventful recovery and final pathology revealed necrotizing aortitis (NA). She is currently undergoing routine surveillance of her descending thoracoabdominal aneurysm. Recent case series indicate that NA is a histologically distinct process, which is associated commonly with development of ascending aneurysm, and although it is most commonly an isolated finding, it may be associated with other vascular abnormalities including stenoses of branch vessels. There is a growing body of literature suggesting that NA represents a distinct clinical entity, associated with the development of ascending aortic aneurysms. Further research is required to determine the optimal follow-up and value of medical therapies.

KEYWORDS: Aorta, surgery, etiology, aortic aneurysms

The term *aortitis* describes inflammatory conditions of the aorta, which can be further subdivided into infectious and noninfectious categories.¹ Infectious

causes of aortitis have classically been linked to pathogens including *Syphilis*, *Salmonella*, *Staphylococcus*, and *Mycobacterium*.² Noninfectious aortitis is much more

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common and associated with the development of aneurysm formation.³ Although descending aneurysms are generally associated with the development of atherosclerosis, cystic medial necrosis, and smoking, ascending aneurysms are more likely to be associated with an inflammatory condition of the aorta.¹ Noninfectious aortitis of the ascending aorta is increasingly found in isolation during resections of otherwise asymptomatic aneurysmal disease.⁴ We present here a case recently managed at our institution.

CASE REPORT

We present the case of a 64-year-old Caucasian woman who presented to our medical center with an incidentally discovered thoracoabdominal aneurysm. She had previously been in good health and had not complained of chest pain or been otherwise symptomatic. At presentation to our clinic, her ascending aorta measured 5 cm at the sinotubular junction (STJ) with dilation of her descending thoracic aorta to 6 cm. Her coronary angiogram was normal. Cross-sectional imaging was notable for thickening of the aortic wall along its length (Figs. 1, 2).

Operative repair of the ascending arch was conducted using an interposition graft. The aortic valve apparatus was normal, and the proximal anastomosis was created at the STJ. Intraoperatively, an inflammatory obliteration of the aortopulmonary window was noted. It was not possible to completely excise the posterior aortic wall, and a small cuff of tissue was left attached to the pulmonary trunk.

Our patient had an uneventful recovery and was discharged home on postoperative day 5. Final pathology revealed necrotizing aortitis (NA) characterized by an intense inflammatory reaction. Giemsa stain and Fite's acid fast stain were negative for the presence of organisms (Fig. 3A, B).

The decision was made not to start supplemental glucocorticoid therapy given the absence of other signs of a serological cause for her vasculitis. She is currently

undergoing routine surveillance of her descending thoracoabdominal aneurysm, which will also likely need repair in the future.

DISCUSSION

Incidental discovery of noninfectious aortitis is made in up to 10% of specimens resected during the repair of aortic aneurysms.^{2,5} Noninfectious aortitis encompasses a spectrum of disease including Takayasu's arteritis, giant cell arteritis, and temporal arteritis, conditions that also generally involve multiple organ beds. However, there is increased awareness of isolated aortitis as an entity, which may not manifest with other signs or systemic disease.^{1,4}

There are distinct pathological findings in cases of noninfectious aortitis. Burke and colleagues³ have proposed a classification based on histological appearance. Following detailed examination of a series of 52 cases of ascending aneurysm, two different patterns were identified. NA was characterized by zonal medial laminar necrosis rimmed by giant cells. Healed areas were common with extracellular accumulation of proteoglycans giving the appearance of medial degeneration. Non-necrotizing aortitis was characterized by the absence of necrosis with diffuse medial inflammation. NA was concluded to be largely an isolated process and only infrequently associated with other seropositive conditions.

When an isolated inflammatory condition of the aorta is identified, a full workup is warranted. Once infectious causes have been out ruled, it is prudent to further evaluate the patient for serological issues and may warrant involvement of a rheumatologist. At a baseline, evaluation of C-reactive protein levels and erythrocyte sedimentation rate should be performed and, although not useful in determining the etiology, may be helpful in monitoring response to treatment in the event that a systemic inflammatory/autoimmune condition is diagnosed. Associated conditions may be found in up to 49% of patients with incidentally discovered aortitis.²

There are some important considerations in the surgical management of the ascending aorta in the setting of aortitis. It is important to recognize that this inflammatory condition may be associated with both ostial coronary stenosis and branch vessel stenosis.⁶ It is also associated with the development of severe aortic regurgitation.⁷ In this case, the valve leaflets are usually normal⁵ and valvular insufficiency may simply be due to sinotubular enlargement, which may be corrected by interposition of a well-sized graft conduit. That being said, careful follow-up is warranted as further aneurysmal dilation of the root may occur in a small proportion of patients due to progression of disease.

Figure 1 Axial section of aortic root demonstrating enlargement at the level of the sinotubular junction to 5.3 cm.

Figure 2 Sagittal section of aorta demonstrating thickening of the aortic wall, most prominent in the descending aorta. The soft tissue thickening is most notable as it relates to branch vessels (superior mesenteric artery shown).

Figure 3 Sections stained with hematoxylin and eosin (A) and elastic trichrome (B) show a striking aortitis characterized by extensive, bland, zonal medial laminar necrosis containing elastic lamellar remnants. These are surrounded by a granulomatous giant cell reaction. Other areas show medial degeneration with excess extracellular ground substance/matrix. Atherosclerotic and fibrotic changes are present in the intima, and the adventitia shows a mixed lymphoplasmacytic infiltrate without endarteritis obliterans or granulomas.

Whether the finding of an isolated arteritis should be treated postoperatively with glucocorticoids remains a matter for debate. There is some evidence to suggest that medical management may affect the progression of disease and its manifestation in other vascular beds. In a series of 52 patients followed by Rojo-Leyva and colleagues,⁸ there was a 17% occurrence of new aneurysms in the 3-year follow-up period in patients not treated. More recently, however, Miller and colleagues⁴ followed 21 such patients over a mean period of 81 months with only 1 of 19 not treated with steroids developing new disease. Therefore, the risk incurred in prescribing steroids with regard to wound healing and other complications may outweigh the potential theoretical benefit of such therapy. Nevertheless, diligent screening and follow-up are indicated in all patients found to have a noninfectious aortitis at the time of surgery. Regular serial cross-sectional imaging should be performed to evaluate for progression of disease or development of new aneurysms.

CONCLUSION

Although there is increasing awareness of necrotizing or noninfectious aortitis, it remains a poorly understood entity with optimal medical management still unclear. Further studies are necessary, especially with regard to the role of steroids and other immunomodulation therapy.

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