Respecting Symptoms in Thoracic Aortic Aneurysm Management: A Case of Symptomatic Necrotizing Granulomatous Aortitis

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Abstract

A 41-year-old woman presented with chest pain of unclear etiology in the setting of a mildly dilated ascending aorta. Computed tomography angiography showed an aorta with an irregular contour and an aneurysm of 4.5 cm. There was no radiographic evidence of rupture or dissection. The patient was taken to the operating room and was found to have severe aortitis with marked localized wall thinning at imminent risk of aortic rupture. Aortic pathology demonstrated necrotizing granulomas of noninfectious etiology. This case illustrates the importance of respecting symptoms in surgical decision making for thoracic aortic aneurysms that may not meet standard interventional criteria.

Keywords

- aortic aneurysm
- aortic operation
- aortic pathology

Idiopathic aortitis is an increasingly recognized entity, yet one that remains largely underappreciated.^{1,2} Isolated idiopathic aortitis is typically diagnosed when secondary sequelae of the vasculitis, such as an aneurysm or dissection, prompt surgical intervention, and subsequent intraoperative biopsy.³ Here, we present the case of a patient with an ascending aortic aneurysm who harbored a dangerous idiopathic aortitis that was not discovered until she was operated on based on her vague pain symptoms. This case underscores the need to consider aneurysm resection in symptomatic patients even if they do not meet standard dimensional criteria for intervention (5.5 cm for the ascending aorta and 6.0 cm for the descending aorta).

Case Presentation

A 41-year-old woman (169 cm tall, 82 kg) presented to the emergency department of our institution complaining of progressive chest pain for the past week. The pain was located to the right of the sternum and was associated with mid-scapular and right neck pain. Her symptoms were initially

intermittent but had become progressively constant over the preceding week. The pain was not exertional or pleuritic, and she reported no exacerbating factors. She denied any shortness of breath. She had no history of similar symptoms. She was on no medications and did not smoke. Her father had an abdominal aortic aneurysm.

The patient had initially been evaluated by our service 3 years previously, when she was found to have a murmur on routine examination by her primary care physician. Echocardiography demonstrated a trileaflet aortic valve with mild aortic insufficiency and an aneurysmal ascending aorta of 4.3 cm.

Physical examination upon current presentation demonstrated a 2/6 systolic murmur but was otherwise normal. Pulses were symmetric and intact throughout. Electrocardiogram showed sinus rhythm without ischemic changes, and her troponin T and creatine kinase-MB levels were non-elevated. A computed tomography angiogram of the chest revealed that the patient's aorta was irregular in contour and her proximal ascending aorta now measured 4.5 cm in diameter (► **Fig. 1A**). There was no radiographic evidence of

Fig. 1 (A) Computed tomography angiogram of the chest obtained upon presentation to the emergency department demonstrates the aneurysmal ascending aorta with an irregular (noncircular) cross-sectional contour. (B) Intraoperative photo of the inflamed ascending aortic aneurysm (head is toward top of figure). Note the irregular, asymmetric contour of the aorta (arrow).

rupture or dissection. There were no other aneurysmal zones. The patient was admitted to our institution and subsequent cardiac catheterization and aortogram demonstrated no disease in the coronary arteries and a proximal ascending aorta measuring 4.8 cm with effacement of the sinotubular junction and an irregular contour. The arch branch vessels showed no signs of stenosis.

Based on the patient's symptomatology, without a clear nonaortic cause, and the documented presence of a thoracic aortic aneurysm, she was taken to the operating room for aortic root and valve replacement. Intraoperatively, the aortic root and ascending aorta were notably irregular with a moth-eaten appearance (►Fig. 1B). Areas of the aortic wall were markedly thin and translucent such that aortic rupture seemed imminent. Inflammatory changes were noted from the aortic annulus to the base of the innominate artery. Therefore, the patient underwent aortic root replacement with a composite graft (21 mm St. Jude valved conduit, St. Jude Medical, Inc., St. Paul, MN) with coronary button reimplantation. Given the extent of the inflammatory changes including the base of the innominate artery, we also performed a formal arch replacement extending up to the origin of the left carotid artery under deep hypothermic circulatory arrest (18 mm Hemashield vascular graft; MAQUET Cardiovascular LLC, Wayne, NJ). The base of the innominate artery was additionally replaced with a 10 mm Hemashield vascular graft and connected to the 18 mm arch graft via an end-to-side anastomosis. The patient recovered well postoperatively without complications.

Gross pathologic examination found the aorta to be irregular and glistening with multiple yellow plaque-like lesions covering ~50% of the luminal surface. The aortic wall had an uneven thickness on both gross and microscopic analysis, with focal areas showing severe thinning (►Fig. 2A), as noted in the operating room. Necrotizing granulomas with mononuclear epithelioid histiocytes were found throughout the sample (►Fig. 2B, C). Elastic-Verhoeff's von Gieson stain showed heterogeneous destruction of elastic fibers in the aortic wall (►Fig. 2D). Special stains for mycobacteria (acid-

fast bacilli), spirochetes (Warthin–Starry), and fungus (Grocott) were all negative.

Discussion

A final pathologic diagnosis of necrotizing granulomatous inflammation and myxoid degeneration was made. The etiology of this large vessel vasculitis remains unknown. The location and nature of her inflammation (necrotizing and granulomatous) plus the patient's age are suggestive of Takayasu arteritis; however, several clinical and pathologic features warrant an alternative diagnosis. She had none of the clinical symptoms traditionally associated with Takayasu arteritis, including pulselessness, ocular disturbances, neurological deficits, hypertension, or systemic symptoms. In contrast to classical Takayasu arteritis, the aortic wall was noted to be particularly thin, and other vessels did not display luminal narrowing. Upon subsequent clinical workup, the patient was found to have negative purified protein derivative and venereal disease research laboratory tests and a positive antinuclear antibodies test. Erythrocyte sedimentation rate and C-reactive protein tests were within normal limits.

The standard of performing thoracic aortic aneurysm repair based on clinical symptoms, even when standard dimensional interventional criteria are not met, has been previously supported by our team.⁴ In the present report, the patient had idiopathic aortitis with wall thinning that put her at high risk for catastrophic aneurysm rupture. The only warning sign was her atypical chest pain. This case underscores the notion that a symptomatic thoracic aortic aneurysm must warrant consideration for immediate surgical repair.

This case also highlights the notion that our current classification systems for vasculitis do not accommodate all cases and that certain presentations do not neatly fall into specific diagnoses.⁵ The terms "idiopathic aortitis," "nonspecific aortitis," "unclassified aortitis," and "aortitis of obscure etiology" have all been used to describe isolated aortitis that does not fit into previously defined syndromes.⁶ It is possible that a subset of these cases, such as the patient presented here, represents

Fig. 2 (A) Microscopic examination of the aorta (hematoxylin-eosin stain, 40 $\times$) shows the uneven thickness of the aortic wall and focal areas of thinning (arrow). There are also slightly thickened intima and chronic inflammation in the media and adventitia. The labeled portion of the aortic wall (intima, media, and adventitia) represents an area of “normal” thickness. (B) Higher magnification (200 $\times$) shows clusters of epithelioid histiocytes and lymphocytes and (C) areas of focal necrosis. (D) Elastic-Verhoeff’s von Gieson stain (40 $\times$) shows multifocal heterogeneous destruction of elastic fibers, likely contributing to the wall thinning and irregular contour of the patient’s aorta.

atypical presentations of known vasculitides. However, there is mounting evidence that such isolated aortitis may in fact be the most common form of aortitis.^{1,2} While granulomatous inflammation, medial necrosis, and wall thickening have been previously described as part of this condition,⁶ our case of well-formed necrotizing granulomas with wall *thinning* is an unusual presentation, especially in light of the patient’s symptomatology. The aortic wall is a dangerous place to have tissue necrosis. We use this case to document the unusual pathology (necrotizing granulomas) and to emphasize that “symptoms trump size”⁴ in aortic disease.

Conclusion

Acutely symptomatic thoracic aortic aneurysms must be respected. Surgical repair for such aneurysms should be considered even if standard interventional criteria are not met.

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