



Commentary: Aortitis Prognosis: Pathologic Diagnosis Independently Adversely Affects Long-Term Survival

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Aortitis is an uncommon histopathologic diagnosis after thoracic aortic surgery. The clinical presentation of these patients can be highly variable, ranging from completely asymptomatic to dissection or rupture.¹ Cardiothoracic surgeons are increasingly encountering this entity in postoperative surgical specimens.² A more recent consensus statement³ has helped define and classify aortitis. Noninfectious aortitis is mostly caused by either primary large vessel vasculitis including giant cell arteritis and Takayasu arteritis or systemic rheumatological disease. Sometimes aortitis is diagnosed in absence of any clinical features of systemic vasculitis, now defined as clinically isolated aortitis (CIA).³ This can be identified radiologically or in pathologic specimens. When this involves ascending aorta, it is categorized as “Isolated Ascending Aortitis.”

Another study reviewed aortic tissue from 178 patients undergoing elective ascending aortic replacement.⁴ They found that the aortitis patients ($n=26$) had decreased survival at 3 and 5 years. On histopathology, giant cell aortitis was present in the majority of these cases (92%), and lymphoplasmacytic infiltrate was identified in the remainder.

Although this critical study reports a higher rate of aortitis (15%) in patients undergoing ascending aortic surgery than others,^{2,5,6} it confirms many of the results found in earlier literature and provides an insight into the outcomes and natural history of the disease. Most aortitis patients are females, of Caucasian descent and elderly, with age >65 years. As reported previously,^{2,3,5} 73% of patients (19/26) in this report had CIA.

Clifford et al⁵ reviewed 7551 patients who underwent proximal aortic surgery. They identified 196 patients (4%) with noninfectious proximal aortitis. The majority had CIA (66%) at the time of surgery. Importantly, during follow-up of 56 months, 19% of these patients presented with new symptoms and 45% demonstrated new vascular lesions. Of these, 40% of the patients needed 74 additional surgeries, most frequently on distal aorta and branch vessels. This raises important questions,



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Central Message

Diagnosis of aortitis in surgical specimens portends a higher subsequent disease recurrence and declining long-term survival.

whether these patients who were initially categorized as CIA actually represented a heterogeneous aortitis group, and their systemic symptoms were actually subclinical at the time of initial surgery and manifested later on.

Another study from the Mayo clinic picked out 45 cases of active noninfectious aortitis among 513 ascending aortic specimens.⁶ Similarly, the most frequent players in this study were isolated aortitis (47%), giant cell arteritis (31%), Takayasu (14%), rheumatoid (4%), and unclassified (4%). Women were primarily affected.

This report along with the work of others^{2,5-7} helps us understand the implications of aortitis in surgical pathology specimens, particularly in terms of their subsequent care and long-term survival. The authors found a decrement in 3- and 5-year survival of 82% and 72%.⁶ Their results are in agreement with the 55% estimated survival in 8 years as reported by the Cleveland Clinic study.² Given the low long-term survival, there is a need to develop multi-institutional aortitis registries with long-term follow-up on these patients to better understand their cause of death and how can we modify the natural history of the disease.

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Svensson et al² have demonstrated excellent operative outcomes in patients with aortitis, with a low 3.2% mortality (5/156) and 1.9% stroke rate. However, a high rate of new vascular lesions needing subsequent reintervention with declining long-term survival in these patients emphasizes the importance of rigorous and lifelong clinical and radiological surveillance. This also raises an important question that should we perform more extensive initial procedures, such as prophylactic aortic root, or total arch replacement with or without frozen elephant trunk, on these patients at the time of ascending aortic replacement? However, the early risks of extensive aortic replacement should be carefully balanced with the late risks of subsequent redo sternotomy or distal aortic repair.

Furthermore, it remains uncertain as to which subset of aortitis patients would benefit from prophylactic anti-inflammatory or novel immunosuppressive therapies after complete surgical excision.

This particular study is small and retrospective in nature. It represents the tip of the iceberg in terms of our understanding of natural history of aortitis. In addition, 59 specimens were not included due to incompleteness of the data set. While positron emission tomography scan is emerging as an important tool in the management of patients with aortitis,⁷ an analysis of only 6 patients is clinically more interesting than it is statistically helpful. Positron emission tomography can be a highly useful adjunct to the initial diagnosis and follow-up of these patients, particularly to determine the response to anti-inflammatory therapy.

Surgeons must be vigilant in the analysis of pathologic specimens, so that a multidisciplinary plan of care is developed for

these patients including cardioaortic surgeons, rheumatologists, radiologists, vascular surgeons, cardiologists, and primary care physicians. It is clear that these patients cannot be lost to follow-up, or additional mortality/morbidity may ensue. We applaud the authors for shedding some light on a topic that needs further understanding.

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