Noninfectious aortitis and ascending aneurysms: The tip of the iceberg

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Disclosures: Author has nothing to disclose with regard to commercial support.

Received for publication July 3, 2015; accepted for publication July 6, 2015; available ahead of print Aug 1, 2015.

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J Thorac Cardiovasc Surg 2015;150:595-6

0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2015.07.011>

You are operating on a 66-year-old man with a 6.2-cm ascending aneurysm that extends into the arch. He has hypertension, hypercholesterolemia, peripheral vascular disease, and is a former smoker with 2-vessel coronary disease and severe aortic insufficiency with preserved ventricular function. The aorta appears atherosclerotic, consistent with the patient's age and comorbid conditions. You replace the aorta, including the proximal arch, and perform aortic valve replacement and a double bypass operation. The patient does well and is discharged uneventfully. In preparation for a postoperative visit you review the pathology report and are surprised by a diagnosis of noninfectious aortitis. Now what?

In this issue of the *Journal*, Fujimoto and colleagues¹ provide long-term follow-up data on the Mayo Clinic's 57-year experience with repairing ascending aneurysms in patients with noninfectious aortitis. Just as the visible tip gives little indication of the size, shape, and danger of a submerged iceberg, so is the superficial similarity between a patient with an atherosclerotic aneurysm and aneurysm secondary to aortitis. Patients with aortitis are more likely to be women with larger aneurysms but are otherwise indistinguishable from their counterparts with atherosclerosis. Although a small percentage may have a history of temporal arteritis, Takayasu's disease, or an autoimmune disorder, <20% have ever had an inflammatory illness. Acute-phase reactants such as erythrocyte sedimentation rate and C-reactive protein level are rarely elevated. Whereas nearly 10% of ascending aneurysms are inflammatory in nature, the gross appearance at the time of surgery is quite similar to a generic atherosclerotic aneurysm. Less-extensive aneurysmectomy may seem justified. However, 72% of patients with aortitis have significant vascular abnormalities beyond the ascending aorta.² The diffuse nature of this inflammatory process led these experienced surgeons to perform arch reconstruction in nearly half of their patients, a 40% increase in the incidence of significant arch dilation when compared to patients with atherosclerotic aneurysms. Not surprisingly, 28% of these arch resections required either concomitant descending aneurysm repair or an

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Central Message

Patients with aneurysms secondary to noninfectious aortitis need regular systemic vascular evaluation and lifelong follow-up imaging

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elephant trunk as a sensible first-stage procedure. One-fifth ultimately required additional surgery on the downstream aorta.

Although aortic valve procedures were less common in patients with aortitis, the long-term durability of a valve preservation strategy could also be questioned. A previous experience in patients with giant cell aortitis failed to reveal pathologic involvement of the aortic valve.³ However, 22% of patients undergoing a valve-sparing operation in this report required aortic valve replacement within 10 years of surgery.¹ Unfortunately, the pathology of those valves excised at the time of reoperation was not reported.

Thankfully, the iceberg will not necessarily sink the boat. Despite the need for more extensive aortic replacement the surgical risk was low and not greater than in patients requiring aneurysm repair for other pathologic entities. The long-term survival was also similar to patients with atherosclerotic aneurysm. Once aortitis is confirmed, patients should be referred to a rheumatologist for a comprehensive evaluation for systemic disease. Yearly surveillance of the entire vascular system is mandatory to identify additional aneurysms and/or occlusive disease. In those rare cases of acute aortitis, treatment with anti-inflammatory agents is indicated and longer-term immunosuppression may be warranted. However, patients should be counseled

that the efficacy of medical therapy in preventing disease progression is uncertain.⁴

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