Symptoms Matter: A Symptomatic but Radiographically Elusive Ascending Aortic Dissection

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Abstract

Keywords

- ▶ thoracic aortic aneurysm
- ▶ aortic dissection
- ▶ aortic operation
- ▶ symptomatic aneurysm
- ▶ intervention criteria
- ▶ genetic testing for thoracic aortic aneurysms
- ▶ MYLK

Symptoms attributable to a thoracic aortic aneurysm (TAA) are a separate indication for prophylactic repair, irrespective of aortic size. We present the case of a 56-year-old female with a history of a thoracic ascending aortic aneurysm (TAAA) and four other heart and arch vessel abnormalities who presented to us with chest pain radiating to her back. Computed Tomography and echocardiography showed no evidence of a dissection and revealed a maximal ascending aortic diameter of 4.2 cm. The patient subsequently underwent root-sparing ascending aortic and hemiarch replacement due to her threatening symptomatology. A focal dissection was discovered intraoperatively, resembling a similar case previously reported by our team.

The risk of catastrophic complications (rupture, dissection, and death) in thoracic aortic aneurysm (TAA) patients increases with increasing aortic diameter.^{1,2} However, aortic size is not the sole determinant for prophylactic surgical repair. The traditional 5.5 cm threshold for intervention pertains only to asymptomatic thoracic ascending aortic aneurysm (TAAA) patients.³ Aortic size becomes irrelevant in the presence of symptoms attributable to a TAAA. This case demonstrates that symptoms should be recognized as a distinct indication for preemptive operative repair, and may even portend the presence of dissections not amenable to detection by imaging modalities.

Case Description

A 56-year-old woman (5 feet 4 inches tall, weighing 147 lbs) presented with sharp, excruciating chest pain radiating to her upper and middle back, combined with a feeling of

“impending doom”. The patient’s medical history was significant for five abnormalities of the heart and great vessels: (1) an ascending aortic aneurysm, (2) a bovine aortic arch, (3) an aberrant vertebral artery originating from the aortic arch, (4) an atrial septal aneurysm, and (5) an apical left ventricular wall motion abnormality due to stress cardiomyopathy she had suffered 7 years ago. The patient reported having experienced multiple similar but less severe episodes of chest pain and shortness of breath over the past years. One year before this episode, she experienced left calf pain and dyspnea but venous duplex of the left lower extremity and chest computed tomography (CT) were negative for deep vein thrombosis and pulmonary embolism, respectively. The patient’s past medical history was significant for takotsubo cardiomyopathy, breast cancer (*BRCA2* gene carrier, treated with double simple mastectomy, and chemotherapy), and postoperative hypothyroidism after total thyroidectomy (under treatment with levothyroxine). She

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also underwent spinal fusion and was a former smoker for 25 years.

There was no evidence of cardiac anomalies or aneurysm disease in the patient's family. However, her father had coronary artery disease and underwent triple coronary artery bypass grafting.

Her physical examination was normal. The electrocardiogram and cardiac enzymes were negative for myocardial ischemia, and cardiac catheterization showed no stenotic lesions in the coronary arteries. Echocardiography revealed mild dilatation of the ascending aorta without dissection. The interatrial septum was hypermobile but without an interatrial shunt. The systolic function of the left and right ventricle was normal and the aortic valve was functioning well. There was also mild tricuspid and pulmonary regurgitation. A CT angiography showed a stable thoracic aortic aneurysm with a maximal diameter of 4.2 cm and no evidence of dissection, as well as mild atherosclerotic disease of the thoracic aorta and moderate atherosclerotic calcification of the abdominal aorta (►Videos 1 and 2). The aortic arch appeared elongated with a minor buckle where it joined the descending aorta, another indication of aortic disease (elongation) and a possible risk factor for dissection.⁴

Video 1

Chest CTA (axial view). Red arrow: isolated vertebral artery, green arrow: bovine arch. CTA, computed tomography angiography. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/doi/10-1055-s-0038-1675849>.

Video 2

Chest CTA (coronal view). Bovine arch (large red arrow), isolated vertebral artery (small red arrow). CTA, computed tomography angiography. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/doi/10-1055-s-0038-1675849>.

Although no aortic dissection was appreciated on the imaging studies, after consideration, the patient was scheduled for semielective surgical correction because of her recurrent symptoms. A root-sparing ascending aortic replacement (22 mm Hemashield graft [Maquet Cardiovascular LLC, Wayne, NJ]) and hemiarch resection under deep hypothermic circulatory arrest (DHCA) was performed. After opening the aorta, we came across three significant findings. First, there were adhesions between the right lateral aspect

Fig. 1 Intraoperative photo of the inflamed ascending aorta. The arrow shows the localized dissection, undetected by echocardiogram and computed tomography.

of the ascending aorta and the superior vena cava and pericardium, indicating past aneurysmal 'activity'. Second, a focal dissection was present just above the sinotubular junction, approximately 1 cm to the right of and cephalad to the left coronary ostium (►Fig. 1). The dissection seemed to have aborted and stopped by the time of the operation. Third, when we transected the ascending aorta distally, we noted that the aortic wall was thickened, edematous, and waterlogged, indicating severe inflammation. DHCA time was 28 minutes, the aortic cross-clamp time was 57 minutes, and total extracorporeal circulation time was 117 minutes. The patient's postoperative course was unremarkable and she was discharged on postoperative day 4.

Discussion

The decision for surgical intervention in this patient was based solely on her symptoms which were severe (she described the chest pain as "excruciating", sharp like "a red-hot poker") and frequent, as the pain led her to the emergency room on multiple occasions. Arch anomalies found on the patient's imaging studies, such as a bovine arch and an aberrant vertebral artery, are also associated with thoracic aortic disease.⁵ Despite the moderate dilatation of her aorta, of approximately 4.2 to 4.3 cm, intraoperative findings included a dissection that had terminated for the time-being but was nonetheless capable of future activity.

We previously reported the case of a 43-year-old woman who presented with chest pain and had a strong family history of aortic dissections.⁶ The patient also described episodes of chest pain that were similar in character to the ones of the present patient. The maximal diameter of her ascending aorta was 4.0 cm as measured by echocardiography, and neither echocardiography nor MRI had revealed any dissection. However, during surgical repair, a subacute focal dissection was discovered, very similar to the one

encountered in the current case. The patient underwent aortic root replacement with a composite graft with coronary button reimplantation.

In contrast to the current case, the previous patient had a significant family history of aortic aneurysms and dissections. In fact, genetic testing was performed and a heterozygous mutation was identified in the *MYLK* gene, p.S1759P (TCC > CCC). The *MYLK* gene encodes myosin light chain kinase (MLCK) which is activated by binding to a calcium/calmodulin (Ca/CaM) complex and leads to phosphorylation of the regulatory light chain of smooth and nonmuscle myosin II. In this way, MLCK determines the contractile function of smooth muscle cells in the ascending aorta. P.S1759P is a missense mutation that blocks MLCK from binding to CaM, inhibiting its activation and causing smooth muscle contractile dysfunction. P.S1759P has been identified as a causative alteration for familial type ascending aortic aneurysm disease which is inherited in an autosomal dominant manner. In affected families, like our patient's, dissections occur when little or no aortic dilatation is present.⁷ Therefore, genetic screening and early surgical repair is crucial in patients with a family history of TAAA.

The present case emphasizes that symptomatic thoracic aortic aneurysms should be resected regardless of size. The cause of the symptoms in these patients is unknown. However, the fact that a covert dissection was found in both our cases justifies the hypothesis of an aortic origin of symptoms. If these patients had not been operated on, these minor dissections would have probably expanded and progressed to acute type A dissections. As our team has stated before,^{6,8} the size criterion of 5.5 cm for the ascending aorta applies only to asymptomatic patients. This highlights the need for early preemptive surgical intervention in symptomatic

patients with thoracic aortic aneurysms. Once again, regarding thoracic aortic aneurysms, "symptoms and family history trump size."

Conflict of Interest

The authors report no conflict of interest related to this work.

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