Classification of Aortic Pathologies

A brief review of thoracic aortic aneurysms and acute aortic syndrome, including classic aortic dissection, intramural hematoma, and penetrating aortic ulcer.

BY SHARIF H. ELLOZY, MD

Physicians have known about diseases of the thoracic aorta for centuries. One of the earliest pathologic descriptions of a ruptured acute aortic dissection was by Frank Nicholls, who performed the autopsy of King George II of England. "In the trunk of the aorta, we found a transverse sissure on its inner side, about an inch and a half long, through which some blood had recently passed, under its external coat, and formed an elevated ecchymosis. This appearance showed the true state of an incipient aneurysm of the aorta."[1] Since then, elucidation of the pathophysiology, advances in imaging, and the establishment of clinical databases such as the International Registry of Acute Aortic Dissection (IRAD) have enhanced our understanding of the disease processes affecting the thoracic aorta. This article describes the classification of acute and chronic diseases of the thoracic aorta.

THORACIC AORTIC ANEURYSM

Thoracic aortic aneurysms (TAAs) are typically described according to their location. Approximately 60% involve the ascending aorta and 40% involve the descending aorta.[2] The prevalence at autopsy is roughly 3%, with a 2:1 male preponderance. The majority of aneurysms are fusiform, although saccular aneurysms can arise, particularly in the setting of a penetrating aortic ulcer (PAU). The most common predisposing factors include systemic hypertension, atherosclerosis, and chronic obstructive pulmonary disease. Genetically mediated connective tissue disorders such as Marfan syndrome, Loeys-Dietz syndrome, and Ehlers-Danlos syndrome type IV can commonly lead to thoracic aneurysm formation.

Histologically, most aneurysms are characterized by medial degeneration. The majority of patients are incidentally diagnosed on thoracic imaging, as only 5% of patients are symptomatic on presentation.[2]

Transthoracic echocardiography offers excellent visualization of the root and ascending aorta but is limited in its ability to evaluate the descending thoracic aorta. Transesophageal echocardiography can visualize more of the distal aorta, but there are windows that are obscured by the bronchi, and the test is semi-invasive. The most common imaging modality to evaluate the entirety of the thoracic aorta is CTA. It is readily available, noninvasive, and allows for rapid, high-resolution imaging of the entire aorta and branch vessels. Although generally safe, the main risk of CTA is the associated ionizing radiation and nephrotoxic contrast. MRA is another useful imaging modality in the evaluation of TAA. In younger patients, ionizing radiation can be avoided, and the contrast agent is not nephrotoxic. However, MRA cannot be performed in patients with implantable pacemakers, and the use of gadolinium is associated with the risk of nephrogenic systemic fibrosis in patients with renal insufficiency. Generally, the presence of symptoms, maximal diameter, and rate of growth are used as criteria for TAA repair.

ACUTE AORTIC SYNDROME

In contrast to TAAs, which are typically asymptomatic and chronic at diagnosis, acute aortic syndrome (AAS), a description coined in 2001 by Vilacosta et al, is an umbrella term encompassing three overlapping clinical entities: classic aortic dissection, intramural hematoma, and PAU.[3] The clinical profile of these patients varies, but they often have similar presenting complaints. These diagnoses tend to be dynamic, in that a patient with a PAU or intramural hematoma can progress to dissection. All three clinical entities can cause similar symptoms, although malperfusion is more likely with dissection.

In general, the classification of these entities is based on their anatomic location and duration of symptoms. Patients are considered to be in the acute phase if symptoms have been present for < 14 days. The acute phase is associated with the highest rates of morbidity and mortality. AAS is also classified anatomically based on the segment of the aorta that is involved. The original DeBakey classification was applied to classic dissections and involved three types: type I involved both the ascending and descending thoracic aorta; type II involved only the ascending aorta; and type III was limited to the descending thoracic aorta. The more widely used Stanford classification is based solely on the location of disease relative to the left subclavian artery. Stanford type A dissection involves the arch or ascending aorta (corresponding to DeBakey type I or type II), while Stanford type B dissection is limited to the descending thoracic aorta (corresponding to DeBakey type III). These classification systems are useful in that they guide treatment.

International Registry of Acute Aortic Dissection

IRAD is a consortium of research centers around the world dedicated to studying the presentation, management, and outcomes of acute aortic dissections.[4] The registry started in 1996 and currently includes 30 referral centers in 11 countries. The University of Michigan Health System is the coordinating center, and data are currently available on 3,800 acute aortic dissection cases. As such, IRAD has significantly advanced the understanding of AASs.

Classic Aortic Dissection

Classic aortic dissection is typically described as starting with a tear in the intima, with propagation of blood into the media and development of a true and false lumen separated by an intimomedial flap. Classic dissection represents 85% to 95% of all patients with AAS. Median age at presentation is 61 years, with a range typically from the fourth to the seventh decade of life. Patients are typically hypertensive (45%–100% of patients), and there is a high prevalence of smoking. Chest and back pain is the most common presenting symptom, seen in 85% of patients. Pulse deficits (30%), hypotension (25%), and syncope (13%) are frequently seen.[5]

Intramural Hematoma

Intramural hematomas are thought to result from rupture of the vasa vasorum into the medial layer of the aortic wall. Unlike classic aortic dissection, there is no identifiable tear in the intima. Intramural hematomas account for anywhere from 5% to 10% of all cases of AAS. They typically occur in patients with significant atherosclerotic disease, and most cases are located in the descending thoracic aortic. Clinically, patients typically present with abrupt chest and/or back pain. Fewer than 10% of intramural hematomas spontaneously regress, and up to 47% may progress to dissection. Patients tend to be slightly older than those with classic dissection (median age, 68 years), with a similar sex distribution (50%–80% are men).[5-7] Although patients may not present with ischemic complications as frequently as in classic aortic dissection, the overall mortality is similar.[8]

Penetrating Aortic Ulcer

PAUs are an outpouching of blood through the internal elastic lamina, typically arising from inflammatory erosion accompanying atherosclerotic plaque. They account for 2% to 7% of cases of AAS.[9] PAUs are often associated with some amount of intramural hematoma and can progress to aortic dissection, pseudoaneurysm formation, or frank rupture. They tend to occur in older patients who have a larger atherosclerotic burden. The majority of PAUs occur in the descending thoracic aorta.

Imaging Considerations in Acute Aortic Syndrome

Imaging considerations in AAS are similar to those for TAAs. CTA is the gold standard, with reported sensitivity and specificity of 100%. Other benefits of CTA in AAS include its ready availability and rapid acquisition time. MRA has a slightly lower sensitivity (95%–100%) and specificity (94%–98%) than CTA. Transesophageal echocardiography is more sensitive and specific than transthoracic echocardiography, but less sensitive and specific than MRA or CTA.[5]

CONCLUSION

Thoracic aortic pathology encompasses a spectrum of acute and chronic disease entities. Our enhanced understanding of the pathophysiology and natural history of these diseases have allowed for improvement in treatment outcomes. Advances in imaging allow for rapid classification and facilitate clinical decision making. Clinicians should have a low threshold to proceed with imaging in a patient with presenting symptoms that point toward AAS. ■

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Sharif H. Ellozy, MD
Associate Professor of Clinical Surgery
Division of Vascular and Endovascular Surgery
Weill Cornell Medical College
Associate Attending Surgeon
New York-Presbyterian/Weill Cornell Medical Center
New York, New York
sharif.ellozy@gmail.com; she9021@med.cornell.edu
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