Pathologies of Progressive Thoracic Aortic Disease

With the natural tendency of the aorta to dilate, there are inherent practical considerations to account for when planning a TEVAR procedure.

BY THOMAS L. FORBES, MD

In many series describing thoracic aortic disease, it is apparent that there are two main differences regarding patient presentation in this anatomic area compared to the abdominal aorta. First, patients are proportionately more inclined to present urgently with acute onset of back pain, as opposed to patients with abdominal aortic pathologies who are more likely to have asymptomatic lesions. Second, thoracic aortic pathologies are more disparate than those in the abdominal aorta and include thoracic aortic aneurysm (TAA), aortic dissection (AD), intramural hematoma (IMH), penetrating aortic ulcer (PAU), and blunt traumatic thoracic aortic injury (BTAI).

The term *acute aortic syndrome*[1,2] has been attributed to the urgent presentation of thoracic aortic pathologies including AD, IMH, and PAU. Once other etiologies such as acute myocardial infarction are excluded, investigations should lead to contrast-enhanced and nonenhanced CT scan of the chest, abdomen, and pelvis. Complete visualization is important to determine the extent of the acute thoracic aortic process and a potential access for endovascular therapies.[3] Once the specific pathologic variant is identified, the appropriate course of treatment can be determined, recognizing the shared pathophysiology of these clinical entities and the understanding that PAU and IMH represent focal manifestations of the classically more extensive AD.[4]

PATHOPHYSIOLOGY

Acquired and genetic conditions can increase susceptibility to these acute thoracic aortic conditions. Of course, the most common pre-existing condition is hypertension, which leads to intimal thickening, fibrosis, and calcification, in turn leading to degradation of the extracellular matrix and eventual disruption of the intima.[1] Genetic conditions can also cause intimal disruption and AD. These conditions include Marfan syndrome, Ehlers-Danlos syndrome, and bicuspid aortic valve, among others.[1]

Although these clinical entities (PAU, IMH, AD) can be viewed as variations of the same disease process, there are clinical features distinct to each. Patients with PAU and IMH tend to be older (the mean age was 74 years in one series[4]) than those with dissections, and most patients are hypertensive. Penetrating ulcers are associated with atherosclerotic disease of the thoracic aorta, whereas AD often occurs in aortas with lesser degrees of calcification. In addition, PAU and IMH tend to affect thoracic aortas of larger diameters than AD.[4] These pathologic distinctions likely reflect differences in the depth of penetration of the aortic wall between PAU, IMH, and AD.

NATURAL HISTORY

The thoracic aorta is a hostile environment for stent grafts, with violent hemodynamic stresses and forces. These forces can lead to eventual failure of thoracic endovascular aneurysm repair (TEVAR) and result in reinterventions. In addition, the thoracic aorta is a dynamic organ that is prone to progressive dilatation, even in the nondiseased state. In a review of more than 1,000 normal thoracic aortas, Hartley et al[5] observed a steady dilatation of approximately 1 cm between teenagers and those in their 80s. This finding was irrespective of sex, race, or the presence of hypertension, pulmonary disease, or diabetes.

With a progressively dilating thoracic aorta subjected to strong hemodynamic forces, and with variable pathologies, it is apparent that thoracic aortic disease is progressive, and when choosing any therapy, either open surgery or TEVAR, one needs to take this into account. This is true for the more diffuse pathologies such as dissections and thoracoabdominal aneurysms, but also for more focal lesions including isolated TAAs, IMHs, penetrating thoracic aortic ulcers, and the most focal of them all, BTAIs.

TAAs

Even the most localized TAAs that are easily treated (at least initially) with TEVAR can require major reinter-

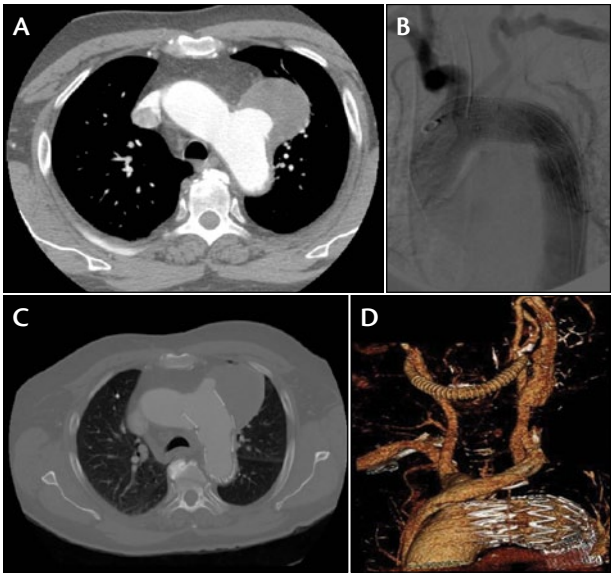

Figure 1. Focal aneurysm of the proximal descending thoracic aorta (A). The initial zone 2 TEVAR was successful, with left subclavian-to-left common carotid artery transposition (B). Two years later, the patient developed a proximal type IA endoleak (C). A carotid-carotid bypass and zone 1 TEVAR were performed (D).

ventions due to the inherent dilatory characteristics of the aorta[5] and the progressive nature of thoracic aortic disease. Figure 1 illustrates a case of a proximal descending TAA that was successfully treated with a zone 2 TEVAR and left subclavian artery–to–left common carotid artery transposition. Two years later, the patient developed a type IA endoleak due to progressive dilatation of the aortic arch and the proximal seal zone. This prompted extension of the TEVAR with a zone 1 proximal landing site and a carotid-carotid bypass.

Any surgeon with even moderate TEVAR experience will recognize this scenario as not being terribly unusual. The published literature is full of reports describing reinterventions following initially successful TEVAR. Our group has published midterm outcomes following TEVAR for all thoracic aortic pathologies.[6] The majority (60%) of these cases were for TAAs. At a median length follow-up of 21 months, 6% of patients developed an endoleak, and 4% of patients, overall, required a TEVAR reintervention.

In a more recent and larger series, the Northwestern group reported their experience with reinterventions following elective TEVAR for TAAs in 83 patients.[7] Overall, 10% of patients required aortic reinterventions at a mean of 32 months following initial TEVAR, indicating the dynamic and progressive nature of thoracic aortic disease. More than half of the reinterventions were for

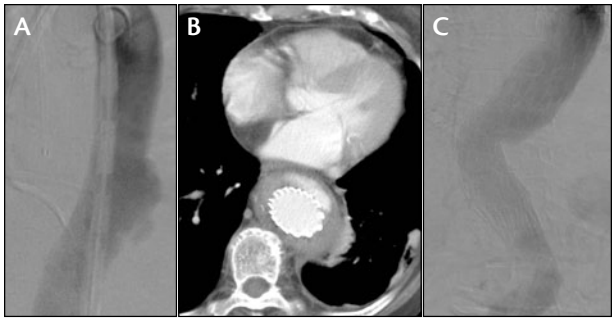

Figure 2. TEVAR for a penetrating aortic ulcer (A). A distal type IB endoleak following initially successful TEVAR for PAU (B). A distal extension was placed for a distal type IB endoleak (C).

type I endoleaks because of progressive aortic dilatation. There was also a trend, although not statistically significant ($P = .052$), for more extensive aneurysms (fusiform) to require secondary interventions compared to those that were more focal (saccular). This information supports the philosophy that all aneurysms of the thoracic aorta represent a panaortic pathology and that no matter how localized the initial endovascular repair, a proportion of these will progress to the extent of requiring reintervention. These issues require attention when planning the initial repair.

IMHs AND PAUs

These lesions are generally more localized than TAAs, but there are numerous examples of the progressive nature of these lesions resulting in further degeneration of the thoracic aorta after initially successful TEVAR. Figure 2 illustrates a case of a PAU with surrounding hematoma that was initially treated with TEVAR with good results. The patient subsequently developed a distal type IB endoleak because of progressive dilatation of the distal descending thoracic aorta. This necessitated a distal extension.

Although AD is widely seen as progressive, less has been known of the progressive nature of more localized pathologies such as IMH and PAU. However, this has recently changed with information provided by the multicenter International Registry of Acute Aortic Dissection (IRAD) that included patients with IMH and PAU. Of the 2,830 patients included in IRAD, 178 had IMH.[8] In-hospital mortality did not differ between IMH of the descending thoracic aorta compared to type B AD (4.4% vs 11.1%; $P = .06$), and mortality at 1 year did not differ either. Very importantly, however, IMH of the descending thoracic aorta was shown to be progressive in nature and resulted in aortic dilatation in more than one-third (39%) of patients.[9] This needs to be considered when planning treatment and the extent of any endovascular

repair, in order to prevent such situations as illustrated in the previous case (Figure 2).

BTAIs

BTAIs are the most localized lesion in the thoracic aorta and most commonly affect patients with normal, nondiseased aortas. However, there is mounting evidence that even these very localized injuries represent a progressive lesion that results in dilatation of the thoracic aorta. In a report from our center involving patients with BTAIs who were treated with TEVAR, we reviewed postoperative CT scans to observe any dilatation at different levels of the thoracic aorta.[10] Although all levels of the thoracic aorta showed some progressive dilatation, as would be expected from natural history data,[5] the segment just distal to the left subclavian artery expanded at a slightly greater rate (0.83 mm per year; P =.025).[10]

Whether this accelerated expansion of the thoracic aorta is a temporary response after BTAI or continues during the longer-term has yet to be determined. How much is due to the BTAI or simply due to the expansile forces of the endograft is also unknown. Recent histologic studies in pigs have illustrated the deleterious effects of endograft oversizing with reduced numbers of muscle and elastic fibers.[11] Longer-term follow-up of these generally young BTAI patients will be essential to determine whether the natural tendency of the normal thoracic aorta to dilate is synergistic with that of the injured aorta and the aorta's histologic response to the presence of an endograft.

CONCLUSION

Although the thoracic aorta is subjected to different pathologies of varying anatomic extents and etiologies, they all have the potential to augment the natural tendency of the aorta to dilate. The natural tendency of the aorta, treated or untreated, is to dilate. This has important ramifications when planning an initial TEVAR procedure in which the length of aortic coverage needs to be carefully determined to minimize the risk of secondary aortic interventions, but not at the expense of neurologic or spinal cord complications. The dynamic nature of the thoracic aorta and the progressive nature of thoracic aortic diseases will continue to challenge vascular specialists involved their treatment. ■

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