Ruptured Descending Aorta Following Acute Type A Dissection Repair in a Patient with Loeys-Dietz Syndrome

Montefiore

Korri S. Hershenhouse MD, Brandon E. Ferrell MD, Joseph J. DeRose Jr. MD
Division of Cardiothoracic and Vascular Surgery, Montefiore-Einstein Medical Center

EINSTEIN
Albert Einstein College of Medicine

BACKGROUND

- Acute dissection complicated by descending aortic rupture is well described in Type B aortic dissection. Open descending aortic repair in the acute phase is highly morbid, with operative mortality exceeding 30%. TEVAR has become the gold standard, with lower mortality rates of approximately 10%.
- Management of acute phase descending aortic rupture following acute type A dissection (ATAAD), is not well described throughout the literature.
- We present a case of aortic rupture following ATAAD repair in a young patient with connective tissue disorder, with unsuitable anatomy for endovascular intervention.

CASE REPORT

- A 28-year-old male presented with ATAAD in the setting of a 4.9 cm root aneurysm. A limited dissection extending from aortic root to innominate artery was seen, with a 1.8 cm arch and 2.2 cm descending aorta. There was no malperfusion.
- Intraoperatively, a primary aortic root tear and severe insufficiency of a trileaflet aortic valve was seen. A new dissection flap was seen propagating through the arch.
- An uncomplicated valve sparing aortic root replacement (VSARR) & hemiarch was performed. The patient was diagnosed post-operatively with Loeys-Dietz syndrome.
- POD 7 – CTA with acute enlargement of the proximal descending to 4 cm. With a small arch for stent grafting and connective tissue disorder, aggressive impulse control and staged re-operative arch and open thoracoabdominal repair was favored.
- POD12 – the patient decompensated with descending aortic rupture and complete true lumen compression distal to the renal arteries. Emergent limited descending thoracic aortic replacement was performed under DHCA.
- Dynamic mesenteric malperfusion ensued post operatively. TEVAR and dissection stenting was performed POD8 with successful re-expansion of the true lumen.
- The remaining course was complicated by ARDS and GI bleed resulting in demise.

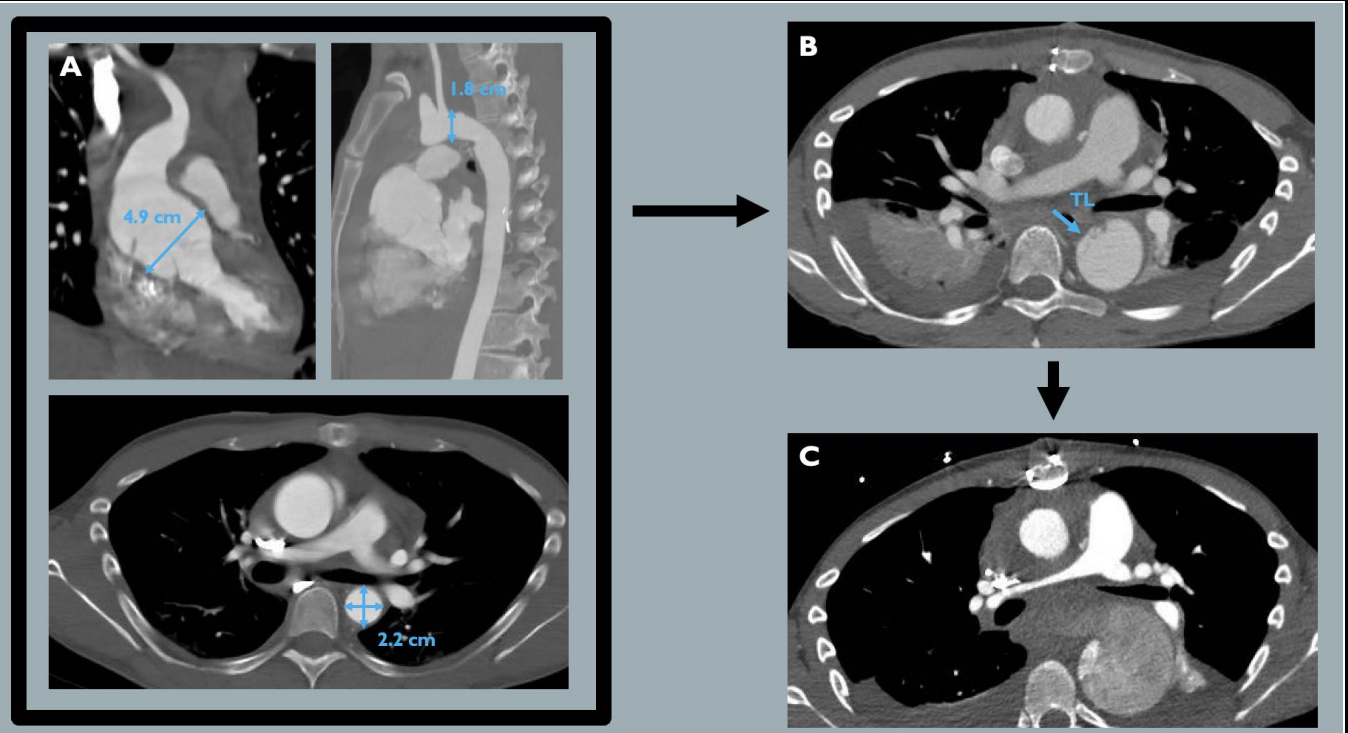

Figure 1: Evolution of descending aortic pathology. A) The presenting ATAAD scan. Limited dissection tear originating from a large root aneurysm, which tapered to normal caliber in the mid-ascending aorta. Small arch and descending aortic caliber. B) POD7 - Acutely enlarged descending aorta with a severely compressed true lumen (TL). C) POD12 - rupturing descending aorta, compressing the posterior trachea.

CONCLUSION

- Descending aortic rupture following ATAAD is a feared complication resulting from persistent pressurization of the false lumen from distal re-entry or fenestrations. The aortic isthmus is prone to aneurysmal degeneration following acute dissection. This patient's arch and distal aortic pathology progressed between his diagnostic CTA and operation, adding challenge to intraoperative decision making. With knowledge of a malignant root aneurysm, small, friable aortic arch, and a distally compressed true lumen, a more aggressive arch reconstruction strategy would have been considered.