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Endovascular repair of a ruptured thoracic aortic dissection with a right sided aortic arch: A case report

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ABSTRACT

BACKGROUND: Emergency treatment of complex aortic pathology is challenging in the setting of a right-sided aortic arch. We report the successful treatment of a ruptured thoracic aortic aneurysm (TAA) in the setting of a Stanford type B aortic dissection (TBAD) and right-sided aortic arch.

PRESENTATION OF CASE: The patient is a 66-year-old male with chronic kidney disease (CKD) admitted with right sided chest pain and hypotension. Computed tomography angiography (CTA) revealed a 5 cm ruptured TAA in the setting of a TBAD and right-sided aortic arch. The TBAD began just distal to the right common carotid artery and involved the origin of the left subclavian artery (SCA). Using a totally percutaneous approach, a conformable Gore® TAG® thoracic endoprosthesis was placed in proximal descending thoracic aorta covering the left SCA. Aside from progression of his pre-existing CKD, the patient had an uneventful recovery. CTA one-month post-procedure revealed a type IB endoleak with degeneration of the distal descending thoracic aorta. To exclude the endoleak, the repair was extended distally using a Medtronic Valiant® thoracic stent graft. The left subclavian artery was subsequently coil embolized to treat an additional retrograde endoleak. The patient has done well with no further evidence of endoleak or aneurysm expansion.

CONCLUSION: Right-sided aortic arch presents challenges in the emergency setting. CTA and post-processing reconstructions are very helpful. While the endoleaks prompted additional interventions, the end result was excellent. This case displays the importance of careful attention to detail and follow-up in these complicated patients.

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1. Introduction

Aortic dissection is one of the most complex conditions encountered in medicine and is associated with significant morbidity and mortality. Aortic dissections are initially classified based on the age of the dissection and on their anatomic location. Computed tomography angiography (CTA) is the imaging study of choice to evaluate this pathology and is vital to the initial diagnosis and classification. Dissections are considered acute, if diagnosed within 2 weeks of onset, or chronic, if diagnosed greater than 2 weeks following initial symptoms. Regarding anatomic location, there are two classification systems used to describe this disease process: The DeBakey system and the Stanford systems. The Stanford classification system divides dissections into those that involve the ascending aorta and those that do not. The DeBakey system is slightly more complicated as it categorizes the dissection based on its origin and extent. In general, aortic dissections that involve the ascending aorta require urgent open surgical repair because of the unacceptable mortality rate associated with medical management [1]. Aortic dissection that do not involving the ascending aorta are further classified as complicated or uncomplicated. Complicated aortic dissections are associated with aortic rupture, malperfusion to vital organs, or refractory pain. In these complicated aortic dissection patients, open surgical or thoracic endovascular aortic repair (TEVAR) is recommended. Uncomplicated dissections are managed most effectively with medical therapy [2]. Although, endovascular treatment of uncomplicated aortic dissections remains an area of controversy that is currently undergoing further investigation [3].

A right-sided aortic arch is an uncommon congenital anomaly with an incidence of 0.1% [4]. A ruptured thoracic aortic aneurysm in the setting of a right sided aortic arch is even rarer with few cases reported [5–10]. The scattered surgical case reports that exist uniformly emphasize the technical challenges associated with each patient's particular anatomy. Unique anatomic findings, associated with a right sided aortic arch, include (1) the pattern of the aortic arch vessels, (2) the location of the descending thoracic aorta

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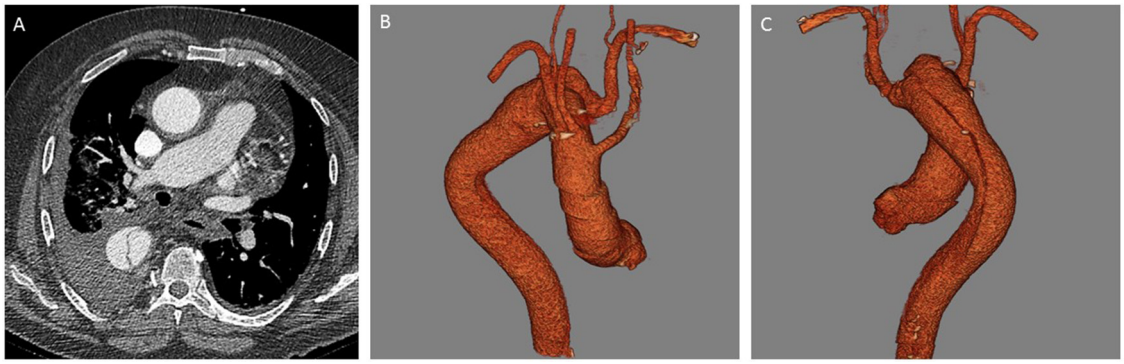

Fig. 1. Pre-operative axial image (A) and three-dimensional volume-rendered images (B [anterior view] & C [posterior view]) from computed tomography angiogram demonstrating an aortic aneurysm confined to the proximal descending thoracic aorta with evidence of contained rupture, an acute type B thoracic aortic dissection and the presence of a right-sided aortic arch.

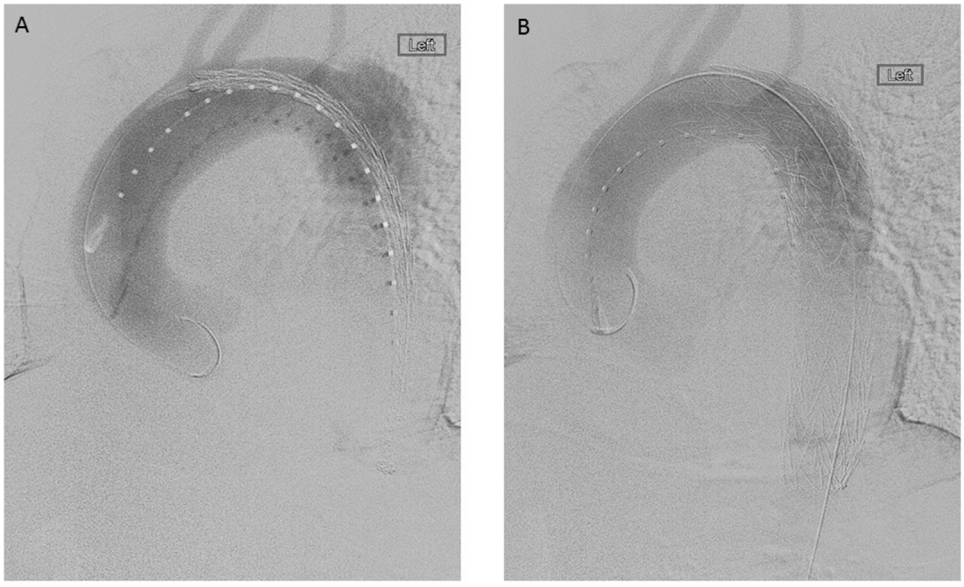

Fig. 2. Digital subtraction angiography in a steep left anterior oblique projection demonstrating the right-sided aortic arch and ruptured aneurysm prior to (A) and following thoracic stent graft exclusion.

(right or left), (3) any anomalous vessels originating from the aortic arch, (4) the relationship of the aorta to the esophagus and trachea and lastly, (5) any associated cardiac anomalies [7]. Endovascular treatment of aneurysms associated with a right sided aortic arch is appealing as it avoids cardiopulmonary bypass and median sternotomy or thoracotomy. We report here the successful treatment of a ruptured thoracic aortic aneurysm (TAA) in the setting of a Stanford type B aortic dissection (TBAD) and right-sided aortic arch.

2. Presentation of case

The patient is a 66-year-old male with stage IV chronic kidney disease (CKD) who was admitted with right sided chest pain and transient hypotension which resolved with fluid resuscitation on arrival. Computed tomography **angiography (CTA)** was performed immediately and revealed a 5 cm aneurysm confined to proximal descending thoracic aorta with evidence of contained rupture. It also demonstrated the presence of an acute type B thoracic aortic dissection (TBAD) and a right-sided aortic arch. The dissection began just distal to the right common carotid artery and involved the origin of the left subclavian artery (SCA). The left common carotid artery arose proximally on the arch and was uninvolved (Fig. 1).

Using a totally percutaneous approach, a 37 mm by 20 cm long conformable Gore® TAG® thoracic endoprosthesis (Flagstaff, Arizona) was placed in proximal descending thoracic aorta covering the left SCA (Fig. 2). A chest tube was inserted to evacuate the right hemothorax. Aside from the institution of hemodialysis due to progression of his pre-existing CKD, the patient had an uneventful recovery. CTA one-month post-procedure revealed a type IB endoleak due to degeneration of the distal descending thoracic aorta. To exclude the endoleak, the repair was extended distally using a repeat percutaneous approach and a 38 mm by 107 mm long Medtronic Valiant® thoracic stent graft (Minneapolis, Minnesota) (Fig. 3). Six months later, the left subclavian artery was coil embolized to treat an additional retrograde (Type II) endoleak (Fig. 4). The patient has done well with no further evidence of endoleak or aneurysm expansion (Fig. 5). Autologous dialysis access was created in his right upper extremity.

Emergency treatment of complex aortic pathology is challenging in the setting of a right-sided aortic arch. CTA, along with post-processing reconstructions, is always helpful. Initial limited stent graft coverage of the thoracic aortic pathology seemed very appropriate until the follow-up CTA revealed a Type IB endoleak. While the endoleak prompted additional intervention, the end result was excellent. This case displays the importance of care-

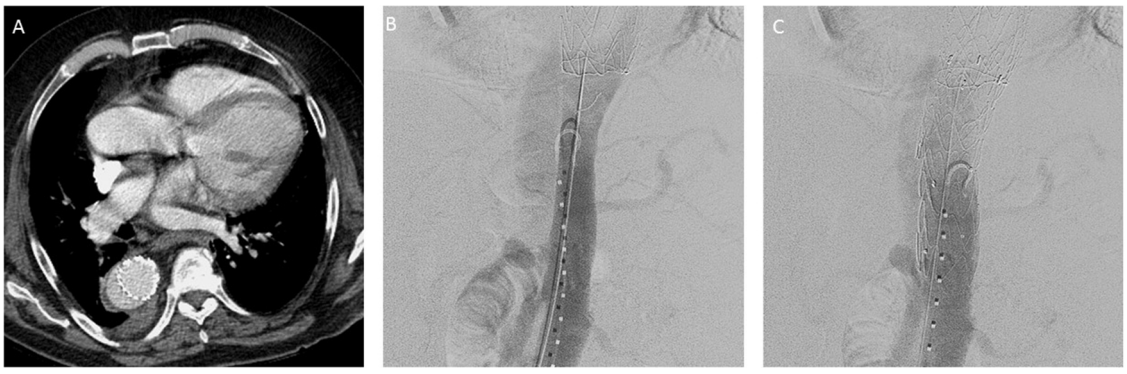

Fig. 3. Axial image from computed tomography angiogram (A) and digital subtraction angiography (DSA) demonstrating a type IB endoleak due to degeneration of the distal descending thoracic aorta. DSA confirming successful stent graft exclusion of the type IB endoleak.

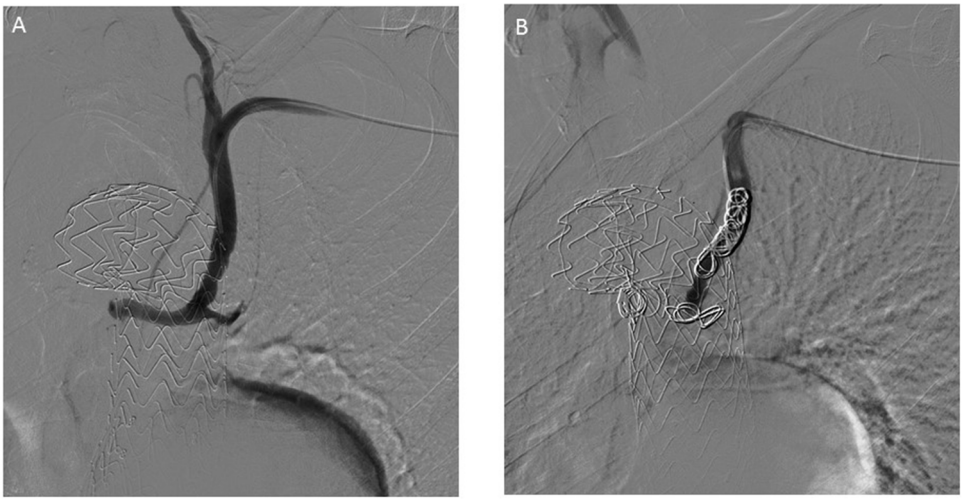

Fig. 4. Digital subtraction angiography demonstrating a retrograde endoleak from the left subclavian artery (A) and successful coil embolization of the left subclavian artery to treat the retrograde endoleak (B).

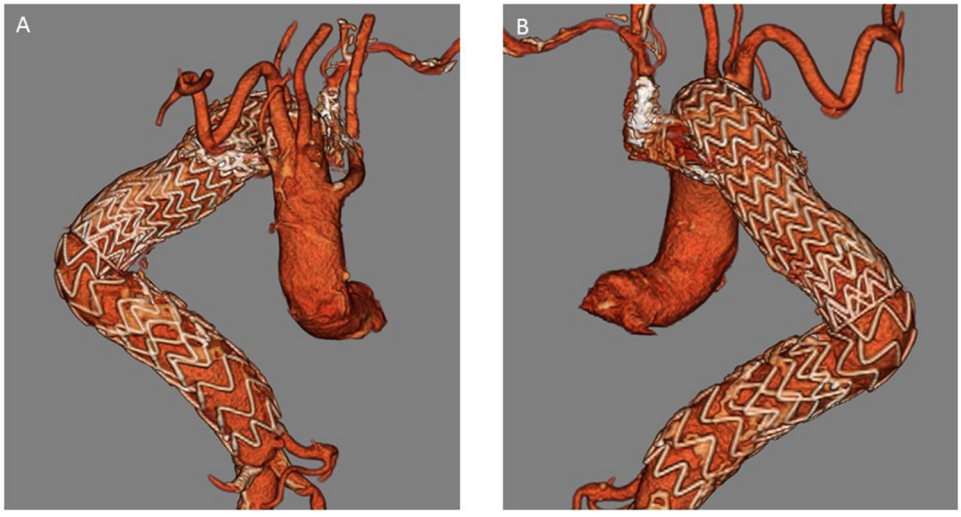

Fig. 5. Three-dimensional volume-rendered images (A [anterior view] & B [posterior view]) from computed tomography angiogram demonstrating a completed successful totally percutaneous repair of a ruptured thoracic aortic dissection with a right sided aortic arch.

ful attention to detail and close follow-up in these complicated patients. Please note this case report follows the CARE guidelines and has also been reported in line with the SCARE criteria [11,12]. The patient has given informed consent for publication of this case report and the Geisinger IRB has approved case presentations.

3. Discussion

Acute aortic dissection presents in different patterns that dictate both the patient's prognosis and the therapies available to the surgeon. Currently, there are two standard classification systems used to describe aortic dissections: The DeBakey and the Stanford

systems. The Stanford classification system divides dissections into two categories: those that involve the ascending aorta (Stanford A dissections) and those that do not (Stanford B dissections). The DeBakey system is slightly more complicated as it categorizes the dissection based on its origin and extent. It is beyond the scope of this case report and will not be discussed further. Acute Stanford type B dissections are further classified as complicated or uncomplicated. Complicated Stanford B dissections include approximately 20% of patients with rupture, malperfusion, or refractory pain. In these complicated patients, open surgical or endovascular therapy is recommended. Uncomplicated Stanford B dissections are managed most effectively with medical therapy [2].

Thoracic endovascular aortic repair (TEVAR) is a relatively new therapy that allows for minimally invasive treatment of thoracic aortic aneurysm and dissections. TEVAR is revolutionizing the care of patients with thoracic aortic pathology as it reduces the considerable morbidity and mortality associated with direct open surgical repair via thoracotomy. Despite the avoidance of thoracotomy and aortic cross clamping, TEVAR is still a major surgical procedure and is associated with serious complications that include stroke and spinal cord ischemia [13]. In this case, the emergent nature of the surgery mandated extending the thoracic stent graft across the origin of the patients left subclavian artery. While this can be necessary in up to 40% of patients, it is associated with increased risks of arm/spinal cord ischemia, stroke and death [14]. An endoleak, defined as blood outside of the stent graft and within the aneurysm sac, occurs following TEVAR in 9–38% of patients [15]. In this case, we were faced with both a Type IB and a Type II endoleak. Type I endoleaks originate from the proximal (Type IA) or distal endograft (Type IB) attachment sites. A Type I endoleak is the most common endoleak found following TEVAR and mandates intervention. In this case, it was treated by extension of the thoracic stent graft distally. Type II endoleaks result from retrograde flow into the aneurysm form aortic side branches. In this case, it arose from the left subclavian artery and was successfully treated with coil embolization. We believe that our case report highlights the need for lifelong imaging surveillance to ensure the success of this minimally invasive treatment.

Ruptured aneurysms and acute complicated dissection occurring in the setting of a right-sided aortic arch are extremely rare. Indeed, a recent review [16] from Barr et al. only identified 99 total patients undergoing surgical or endovascular repair of a right aortic arch aneurysm or dissection. Furthermore, only 12 had a totally endovascular procedure like our patient. Given the rarity of this clinical scenario, the is no current standard of care and many different techniques have been described. We believe that the approach to these patients should depend upon the pathology and the area of the aorta involved. Furthermore, high quality CT angiography, careful pre-operative planning and meticulous follow-up are needed to ensure success when caring for these challenging patients.

4. Conclusion

Emergency treatment of complex aortic pathology is challenging in the setting of a right-sided aortic arch. Computed tomography **angiography**, along with post-processing reconstructions, is always helpful. This case displays the importance of careful attention to detail and meticulous follow-up in these challenging patients.

Conflicts of interest

None.

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Ethical approval

NA.

Consent

Written and signed consent was obtained from the patient.

Authors contribution

JIL – data analysis and interpretation, writing the paper.

JRE – overall responsibility, data analysis and interpretation, writing the paper.

SAF – image preparation and study design.

EJR – data analysis and interpretation, writing the paper.

Guarantor

JRE.

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