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Blunt Traumatic Aortic Injury of Right Aortic Arch in a Patient with an Aberrant Left Subclavian Artery

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Right-sided aortic arch (RAA†) is a rare congenital developmental variant present in about 0.1 percent of the population. This anatomical anomaly is commonly associated with congenital heart disease and complications from compression of mediastinal structures. However, it is unknown if patients are at a higher risk of blunt thoracic aortic injury (BTAI). We report a case of a 20-year-old man admitted to the hospital after being hit by an automobile. Computed tomographic scan revealed an RAA with an aberrant left subclavian artery originating from a Kommerell's diverticulum. A pseudo-aneurysm was also seen along the aortic arch. A diagnosis of blunt traumatic aortic injury was made. The patient was successfully treated with a 26mm Vascutek hybrid stentgraft using the frozen elephant trunk technique.

A literature review of the pathophysiology of BTAI was performed to investigate if patients with right-sided aortic arch are at a higher risk of suffering from BTAI. Results from the review suggest that although theoretically there may be a higher risk of BTAI in RAA patients, the rarity of this condition has prevented large studies to be conducted. Previously reported cases of BTAI in RAA have highlighted the possibility that the aortic isthmus may be anatomically weak and therefore prone to injury. We have explored this possibility by reviewing current literature of the embryological origins of the aortic arch and descending aorta.

INTRODUCTION

Right aortic arch (RAA) is an uncommon anatomical variant that occurs in about 0.1 percent of the population [1]. Two main types are commonly seen: mirror-image branching (Type I) (Figure 1) and aberrant left subclavian artery (LSA) (Type II) (Figure 2). Although extremely rare, there is also a third type, which involves an isolation of the LSA [3]. Type I RAA is commonly associated with congenital cyanotic heart disease, while Type II RAA is often accompanied by a Kommerell's diverticulum, an aneurismal diverticulum that develops at the origin of the LSA and the proximal aspect of descending aorta [4].

Patients with Type II RAA are often asymptomatic and only diagnosed incidentally in adulthood or when complications arise from compression of the mediastinal structures, caused by a growing Kommerell's diverticulum [5]. Current literature regarding RAA focuses on the development of this anatomical variant and the surgical techniques available to manage aortic aneurysmal disease in such patients.

In this paper, we report a patient with a Type II RAA who suffered a blunt thoracic aortic injury (BTAI) caused by a road traffic accident (RTA). Although BTAI is found in only 2 percent of patients who suffer a blunt trauma to the thorax [6], it is ranked the second leading cause of death in individuals aged 4 to 34 [7], with 81 percent of cases caused by automobile collisions [8].

CASE PRESENTATION

A 20-year-old man was admitted to the hospital after being involved in an RTA. On initial examination, the patient was drowsy and only responded to voice. He com-

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†Abbreviations: BTAI, blunt thoracic aortic injury; CPB, cardiopulmonary bypass; CT, computed tomography; LCCA, left common carotid artery; LSA, left subclavian artery; RAA, right aortic arch; RCCA, right common carotid artery; RSA, right subclavian artery; RTA, road traffic accident; TOE, transoesophageal echocardiogram.

Keywords: pseudo-aneurysm, diverticulum, vascular malformation, vascular system injuries

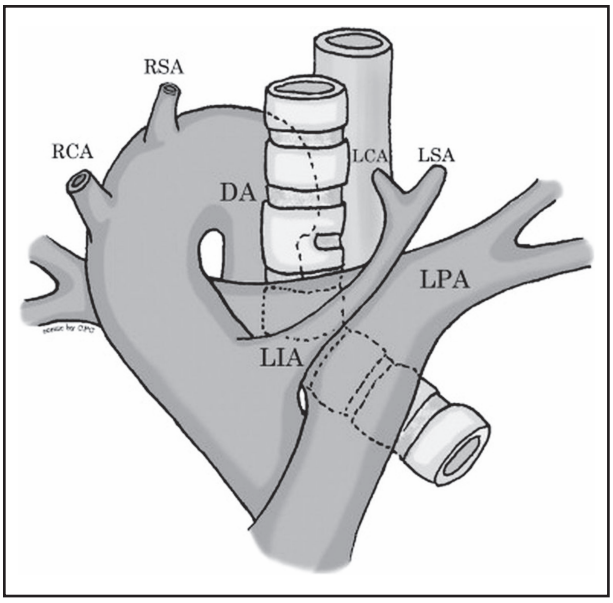

Figure 1. Type I RAA. The ascending aorta arises from the left ventricle and ascends toward the right side. The first branch is the left innominate artery (LIA) that branches to form the left common carotid artery (LCA) and left subclavian artery (LSA). The second branch is the right common carotid artery (RCA) and the third branch is the right subclavian artery (RSA). Adapted from [2].

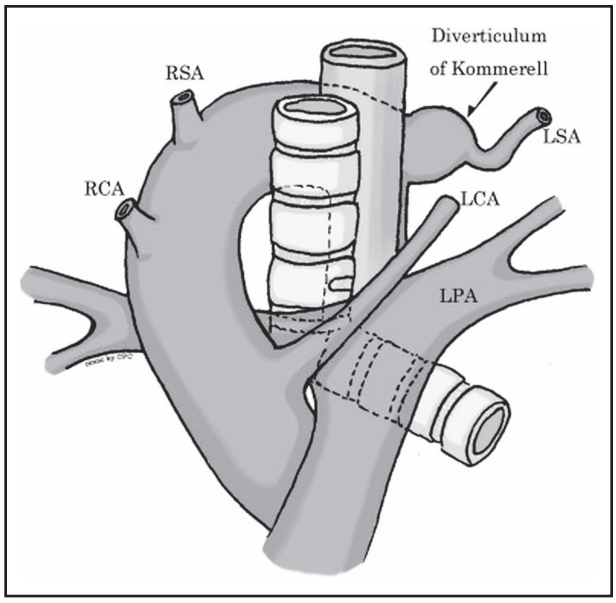

Figure 2. Type II RAA. The ascending aorta ascends from the left ventricle toward the right side and branches off first to form the left common carotid artery (LCA). Its second and third branch is the right common carotid artery (RCA) and the right subclavian artery (RSA) respectively. The left subclavian artery (LSA) branches off at the distal end of the aortic arch, at a retrotracheal position. Adapted from [2].

plained of severe pain arising from the center of his chest that radiated to his back. His respiratory rate was 12 and oxygen saturation was 98 percent on 60 percent oxygen with a non-rebreather mask. His blood pressure was 106/60 mmHg, and his heart rate was regular at 78 beats per min. An urgent whole-body, contrast-enhanced computed tomography (CT) scan was arranged. Based on the patient's presentation, the emergency physicians suspected aortic injury, and hence, a contrast-enhanced CT scan was chosen over non-contrast CT.

The scan revealed a RAA that descends along the right side of the spine before turning left to enter the aortic hiatus at the normal position. An aberrant LSA was also seen arising at the junction of the aortic arch and the descending aorta (Figure 3). The branches of the RAA — proximally to distally — are as follows: left common carotid (LCCA), right common carotid (RCCA), right subclavian artery (RSA), and aberrant LSA (Figure 4).

At the origin of the aberrant LSA, there was a dilatation of the arch of the aorta known as Kommerell's diverticulum. Facing anteriorly, a pseudo-aneurysm arose between the origins of the right and left subclavian artery (Figures 3 and 4). It measured 31.5mm and was compressing the trachea. As the pseudo-aneurysm was anatomically separate from the Kommerell's diverticulum, we believe it likely was caused by the RTA.

An endovascular repair of the pseudo-aneurysm using a covered stent graft was deemed unsuitable due to the proximity of its location to the subclavian arteries, as it may have compromised blood supply to the upper limbs. Instead, a method known as the frozen elephant trunk technique was employed.

A sternotomy was carried out to expose the heart, the ascending aorta, and the aortic arch. A cardiopulmonary bypass (CPB) was established after cannulation of the distal ascending aorta and right atrium. A left ventricular vent was inserted via the right superior pulmonary vein to maintain a dry surgical field and prevent ventricular distension following reperfusion and rewarming — a critical period especially if ventricular function does not return immediately after the release of the aortic cross clamp. A retrograde cardioplegic cannula was inserted through the right atrium to maximize myocardial protection while undergoing hypothermic circulatory arrest. When circulatory arrest commenced at 20°C, the LCCA and RCCA were cannulated to maintain selective antegrade cerebral perfusion. The RSA was anastomosed to a 10mm Hemashield graft, where perfusion of cold blood continued.

Replacement of the damaged aortic arch and re-anastomosis with its tributaries was performed using the frozen elephant trunk technique. A 26mm Vascutek Thoraflex Hybrid stent graft was deployed into the ascending aorta, and its distal endovascular stent was deployed into the proximal descending aorta using a guide wire. A transoesophageal echocardiogram (TOE) was used to verify its position. Distal body circulation recommenced with one of the side arms of the stent graft along with systemic rewarming, while cerebral perfusion remained cool.

During the re-warming period, the LCCA and RCCA were anastomosed to their respective branches of the graft

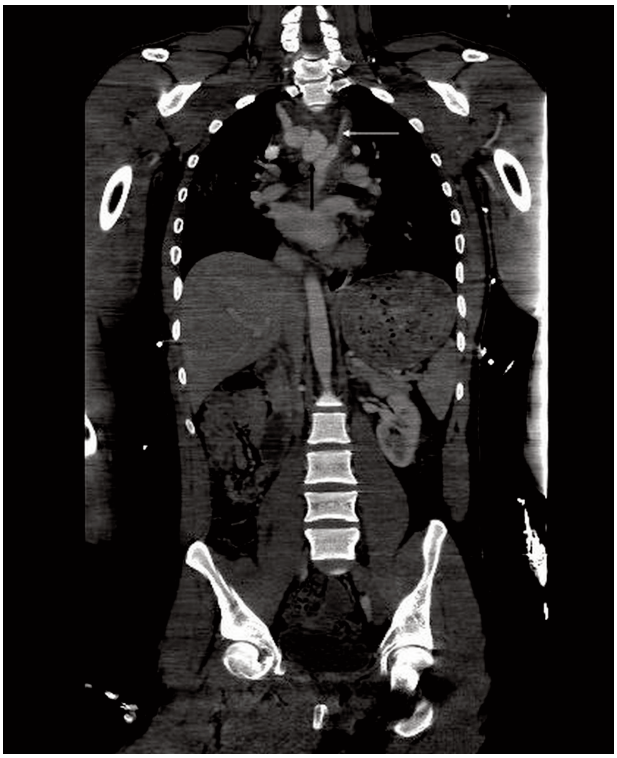

Figure 3. A coronal plane contrast-enhanced CT image. The pseudo-aneurysm (black arrow) and the origin of aberrant LSA (white arrow) are seen.

along its arch, while the RSA was anastomosed to its most proximal side-branch. The aberrant LSA remained untouched. Re-warming of cerebral perfusion was initiated. After reaching a core temperature of 35°C, the patient was weaned off CPB with no complications.

Post-operative TOE showed a well-expanded stent graft with no extravasations (Figure 5). Perfusion to all branches of the aortic arch was also present. The patient was discharged 2 days later.

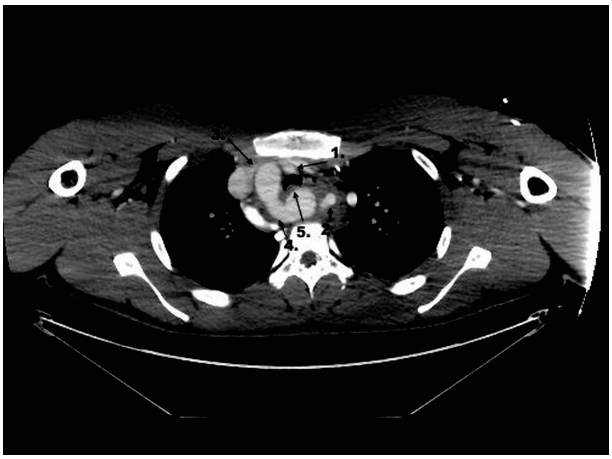

Figure 4. An axial plane contrast-enhanced CT image. The RAA branches off to give the LCCA (1) and aberrant LSA (2). The branching of the RCA (3) and RSA (4) is denoted. The pseudo-aneurysm (5) is also seen compressing the trachea.

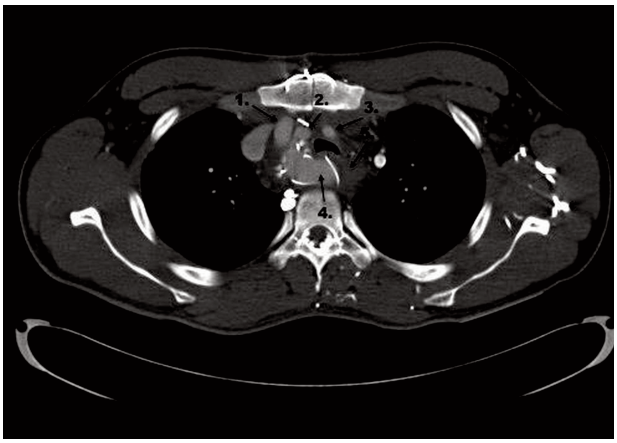

Figure 5. An axial plane contrast-enhanced CT image post-operatively. The RAA branches off first to give the RSA graft (1). The RCCA graft (2) and LCCA graft (3) is also seen. The pseudo-aneurysm(4) has been reduced and the aberrant LSA (5) is not seen.

DISCUSSION

In this reported case, the frozen elephant trunk technique was employed to repair the damage caused by the blunt traumatic injury. This is a variation of the original elephant trunk technique, which was developed in particular to treat extensive aortic disease of the aortic arch and descending aorta in two stages. First, the diseased aortic arch is removed, with the great vessels preserved as an island graft for later anastomosis. A conventional tube graft is invaginated into the proximal descending aorta. The folded end of the graft is sutured to the transected edge of the proximal descending aorta. The proximal end of the graft is then retracted and anastomosed to the transected edge of the proximal aortic arch. The great vessel island graft is then separately sutured to the prosthetic aortic arch. The distal end of the graft remains hanging freely within the descending thoracic aorta until it is utilized in a second surgery to repair the descending aorta [9]. Though initially pioneered in 1983 by Borst et al. [10] to lower the risk of lung, blood vessel, and nerve injury as well as the duration of the aortic cross-clamp times associated with thoracic aortic repair, the elephant trunk technique has undergone decades of modification, refinement, and optimization into various adaptations.

The frozen elephant trunk technique condenses the classical technique into a single step. It is performed with a "hybrid" vascular graft, which is a combination of a conventional tube graft proximally and an endovascular stented graft distally. The distal end, containing a Dacron collar, allows for adaptation of the graft to the aorta — the diameter of which may vary in aneurysmal disease. The Dacron collar, along with the endovascular stenting of the proximal descending aorta, achieves a hemodynamic seal, thereby completing the repair of the descending aorta. The aims of a traditional elephant trunk are thus achieved in a single step without the need for a secondary endovascular or surgical procedure [11].

Table 1. Summary of reported cases of traumatic rupture of right aortic arch.

Authors, year	Age/sex	Location	Relation to aberrant LSA	Surgical approach	Type of repair
Berkoff, 1984	39/F	Greater curvature	Distal	Right thoracotomy	GR
Singh, 1998	20/M	Lesser curvature	Proximal	Median sternotomy	Unknown
Matsumoto, 2006	69/F	Greater curvature	Proximal	Right thoracotomy	GR with ax-ax bypass
Present case	20/M	Greater curvature	Proximal	Median sternotomy	GR

GR, graft replacement; ax-ax bypass, axillo-axillary bypass.

As reviewed by Cinà et al., there are 32 published cases of patients with RAA associated with Kommerell's diverticulum, with 53 percent of these affected patients presenting with rupture or dissection [12]. Most of the current literature revolves around the surgical approach to aneurismal disease in these patients, and there is little published on the pathophysiology behind why these ruptures occur in patients with RAA [13,14].

In 2006, Matsumoto et al. reported a case of traumatic rupture in a patient with Type II RAA, resulting in only three such cases in the published literature [15]. The present case, in addition to the three cases reported by Matsumoto, were all caused by an RTA and associated with an aberrant LSA (Table 1). All four cases also presented a transected pseudo-aneurysm in the region proximal to the origin of the aberrant LSA, where the ligamentum arteriosum attaches. In both right and left aortic arches, this point is anatomically referred to as the aortic isthmus. This alludes to the possibility of the aortic isthmus as an anatomical weak point and hence a common site for BTAI. We propose that the reason for this may lie in its embryological origin.

Over the past 3 decades, fate-mapping studies have shown that the smooth muscle cells in the aortic medial layer have distinct variations in their embryological origins depending on their site [16]. The smooth muscle cells composing the basal aortic root are derived from the secondary heart field. The ascending aorta, the aortic arch, the proximal portion of its cephalic branches along with the ductus arteriosus, as well as remnants of the third, fourth, and sixth aortic arches, all derive their smooth muscle cells from the neural crest [17]. The descending aorta, which is a remnant of the dorsal aorta, obtains its smooth muscle cells from the paraxial mesoderm [16]. It has been suggested that this lineage-derived difference between vascular smooth muscle cells may contribute to the spatiotemporal pattern of vascular diseases. Indeed, studies have proven that adult smooth muscle cells demonstrate varied matrix remodelling responses due to their different origins [18,19,20].

These studies also note that aortic dissections tend to occur at areas where the smooth muscle cells of distinct embryological origins meet. Interestingly, there is growing evidence that the etiology and pathogenesis of aneurismal disease in the ascending aorta and arch of the aorta are very different to that of the descending aorta [18,21]. The ligamentum arteriosum, a remnant of the ductus arteriosus, marks the point of transition between the aortic arch and the descending aorta. We believe that this may explain why the aortic isthmus is an anatomical weak point and prone to injury and dissection. Furthermore, we believe that the presence of a Kommerell's diverticulum in a Type II RAA at this point may further weaken the aorta.

In addition to exploring the embryological origins of the aortic arch and why the aortic isthmus, we also reviewed the literature on the mechanism of BTAI to explore if patients with RAA were indeed at an increased risk of suffering a BTAI.

Several papers have cited that the main mechanism behind BTAI is a sudden stretching force on the aorta. The sudden impact on the aorta causes an abrupt obstruction, leading to a disruption in the laminar flow of blood. Relative to the ascending aorta and the aortic arch, the descending aorta media is less flexible. At this point of transition, where the aortic isthmus exists, there have been studies conducted that have shown this is where the tensile strength of the aorta is weakest. Therefore, any stretching of the vessel wall proximal to the aortic isthmus could increase tension at that point, thus causing a BTAI [22,23,24].

The propagation of a water-hammer effect caused by a blunt injury is another proposed mechanism of BTAI. This occurs when pressure waves created from an abrupt obstruction of blood flow reflect backward along the blood vessel, thereby temporarily increasing stress on the vessel wall. It has been established that an increase in curvature of the aorta can intensify the force of this pressure wave. In RAA patients, there is a more acute curvature of the arch, which intensifies the water-hammer effect, thus potentially causing more damage on the aortic wall [25].

A recent proposed theory also argues that a BTAI is caused by an "osseous pinch" in which entrapment of the aorta between the vertebral column and anterior bony structures, such as the sternum, first ribs and medial clavicles, result in lacerations of the vessel wall. However, it is likely that instead of only one mechanism, it is a collection of the previously mentioned mechanisms that result in a BTAI [26].

These theories suggest that it is an increase in the stress experienced by the aortic wall that ultimately leads to rupture and that this is further exacerbated in a RAA. As we proposed earlier, the aortic isthmus in a patient with Type II RAA could possibly be an anatomical point of weakness based on its embryological origins, as well as by the presence of Kommerell's diverticulum.

In conclusion, based on this information, we propose that patients with Type II RAA may be at an increased risk of BTAI. Unfortunately, due to the rarity of cases for BTAI seen in Type II RAA, it will be difficult to conduct a robust observational study. However, to explore this relationship further, we would like to suggest that investigational studies be conducted, which may be able to link histopathological characteristics of RAA with the biomechanical features of BTAI. An example would be a histological study of RAA in rare patients who do not survive BTAI, as autopsies would be able to reveal the structural characteristics of the aortic wall in such patients, and how the fibrinogen and elastin fibers are positioned at aortic isthmus. This would allow for biomechanical simulation of the stress-strain distribution during traumatic rupture and reveal if the aortic isthmus is truly at the highest risk for BTAI.

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