Aortic arch and frozen elephant trunk repair of a right-sided aortic arch with pseudoaneurysm rupture

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ABSTRACT

A 53-year-old man with a history of vascular ring repair secondary to a right-sided aortic arch with a retroesophageal subclavian artery and ligamentum arteriosum to the descending thoracic aorta presented to our institution with a large aortic pseudoaneurysm of the distal aortic arch. Computed tomography demonstrated a right-sided aortic arch with a 5.8-cm pseudoaneurysm arising from the distal arch with concern for rupture. The patient underwent successful two-stage repair, including a left carotid artery to subclavian artery bypass, followed by total arch replacement with the frozen elephant trunk technique. He recovered well postoperatively, and computed tomography showed complete, successful repair of the pseudoaneurysm. (J Vasc Surg Cases Innov Tech 2023;9:101258.)

Keywords: Aortic arch; Congenital; Frozen elephant trunk; Pseudoaneurysm; Repair

Vascular rings are present in <2% of the population, and, of those cases, a double aortic arch and right aortic arch with left ligamentum arteriosum are the two most common causes.[1] Vascular rings involve the complete encirclement and compression of the trachea and/or esophagus by the aortic arch, its branches, or atretic ligamentous segments. Repair of such a defect was solely performed via thoracotomy until the early 1990s when endovascular repair techniques emerged.[2] Most of the data in long-term follow-up studies discuss persistent tracheal or esophageal compression and concomitant cardiovascular reoperations; however, we highlight a vascular complication after repair of a rare anatomic vascular ring.[3]

CASE REPORT

A 53-year-old man presented with a large aortic pseudoaneurysm of his distal aortic arch. The patient provided written informed consent for the report of his case details and imaging studies. His medical history was significant for a right-sided aortic arch with a retroesophageal left subclavian artery and a ligamentum arteriosum originating from the descending aorta. He underwent repair via left thoracotomy at 6 years of age with division of the ligamentum. It was unknown if he had had a concomitant Kommerell diverticulum resection at that time. He presented with acute-onset right-sided chest pain. A computed tomography angiogram of his chest demonstrated a right-sided aortic arch with a 5.8-cm pseudoaneurysm arising from the distal arch in the area of his previously divided ligamentum (Fig 1 and Supplementary Video 1, online only). The left common carotid, right common carotid, right subclavian, and left subclavian arteries arose independently from the aortic arch.

An open approach from a right thoracotomy was deemed too challenging based on the location of the pseudoaneurysm and technical limitations. Therefore, we elected to perform a two-stage procedure starting with a left carotid artery—subclavian artery bypass with retrograde embolization of the proximal left subclavian artery with an Amplatzer plug (Abbott Cardiovascular). Two days later, we performed total arch replacement with the frozen elephant trunk technique. We approached his aorta through a midline sternotomy. A moderate amount of hemopericardium secondary to contained rupture of the pseudoaneurysm was present, which had not been identified on the preoperative or intraoperative echocardiogram. Central cannulation was performed with dual stage venous cannulation and aortic cannulation proximal to the first branch, the left common carotid artery. A Glidewire (Terumo Interventional Systems) was advanced into the distal arch under transesophageal echocardiographic guidance from a left common femoral artery sheath. He was cooled to deep hypothermia (18°C). His left innominate vein was divided for added exposure.

Once the target temperature was achieved, a cross-clamp was placed in the ascending aorta and the heart arrested with antegrade and retrograde cardioplegia. Deep hypothermic circulatory arrest was begun, and the ascending aorta and aortic arch were opened. The head vessels were transected at their origin from the aortic arch and the stumps oversewn. Bilateral selective antegrade cerebral perfusion was performed by direct cannulation of the right and left common carotid arteries. The right subclavian

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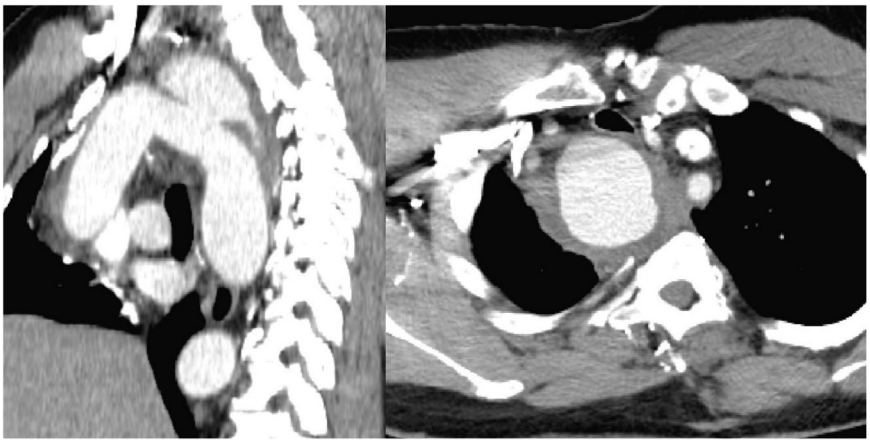

Fig 1. Preoperative computed tomography angiogram of the chest.

artery was difficult to access and of poor tissue quality; therefore, it was ligated. The pseudoaneurysm was noted to be at the origin of the prior ligamentum repair with a contained rupture in the descending thoracic aorta. The previously placed Glidewire was retrieved from the aortic arch, and a Terumo stent graft was positioned over the wire into the proximal descending thoracic aorta. It was deployed just proximal to the ligated left subclavian artery. A commercially available trifurcated woven polyester graft (Terumo Aortic) was used to reconstruct the arch by first anastomosing the distal end to the stent graft and proximal descending thoracic aorta. Lower body perfusion was resumed through a side branch of this graft. The head vessels were anastomosed sequentially, and the proximal anastomosis was performed at the level of the sinotubular junction in the ascending aorta. The cross-clamp was removed, and the patient was weaned from cardiopulmonary bypass. His innominate vein was reconstructed. A right deltopectoral groove incision was made and the right axillary artery identified. A limb of the graft was brought through the right chest and anastomosed to the right axillary artery to reestablish perfusion down the arm. A postoperative angiogram demonstrated exclusion of the pseudoaneurysm, patent reconstruction, and no evidence of an endoleak. Overnight, on postoperative day 0, his left radial arterial line showed a mean arterial pressure 20 points lower than that of his right, and a computed tomography angiogram demonstrated thrombosis of his left common carotid artery graft due to an acute graft angle after chest closure, which required open thrombectomy and reanastomosis of the left carotid graft to the left aorta in a beveled end-to-side fashion on postoperative day 1. However, he recovered well and was discharged to a skilled nursing facility on postoperative day 20. He was subsequently discharged home. Postoperative computed tomography showed successful, complete repair without an endoleak (Fig 2 and Supplementary Video 2, online only).

DISCUSSION

Most children with vascular rings present with symptoms in the first few months of life and require surgery within the first year of life.[4] When the left fourth arch involutes, a right aortic arch is formed with the apex to the right of the trachea.[5] The two variations are a retroesophageal left subclavian artery (65%) and mirror-branching (35%).[6] The subclavian artery originates from the descending aorta and courses to the left behind the esophagus. Patients with a right aortic arch and left ligamentum can have a Kommerell diverticulum at the origin of the left subclavian artery from the descending aorta. Kommerell diverticulum has been repaired successfully endovascularly, which is often the repair strategy.[7,8] In one review of patients with a right-sided aortic arch and Kommerell diverticulum, 6% of the patients presented with rupture and 47% with dissection.[9]

Several operative strategies can be considered depending on the clinical presentation, including endovascular, hybrid, and open. Reviews of the operative strategies for pseudoaneurysm repair after congenital arch repair highlight the efficacy of each technique on a case by case basis.[10-14] Resection of the pseudoaneurysm from a right thoracotomy is technically feasible; however, we believed that given the size and location of the pseudoaneurysm, that approach would have been very challenging. Other repair strategies include total arch replacement, descending thoracic aortic replacement with subclavian artery–carotid artery transposition, and stent grafting with extra-anatomic bypass, which was performed in the present case.[15] Our patient was not a candidate for subclavian artery–carotid artery transposition because a large left thyroid goiter was obscuring the proximal left carotid artery. The frozen elephant trunk approach allowed for exclusion of the descending pseudoaneurysm and treatment of the head vessels in our patient; however, staging with extra-anatomic bypass minimized the circulatory arrest time and addressed the left subclavian aneurysm. Although endovascular stenting with cervical debranching has been described successfully, we did not believe this would

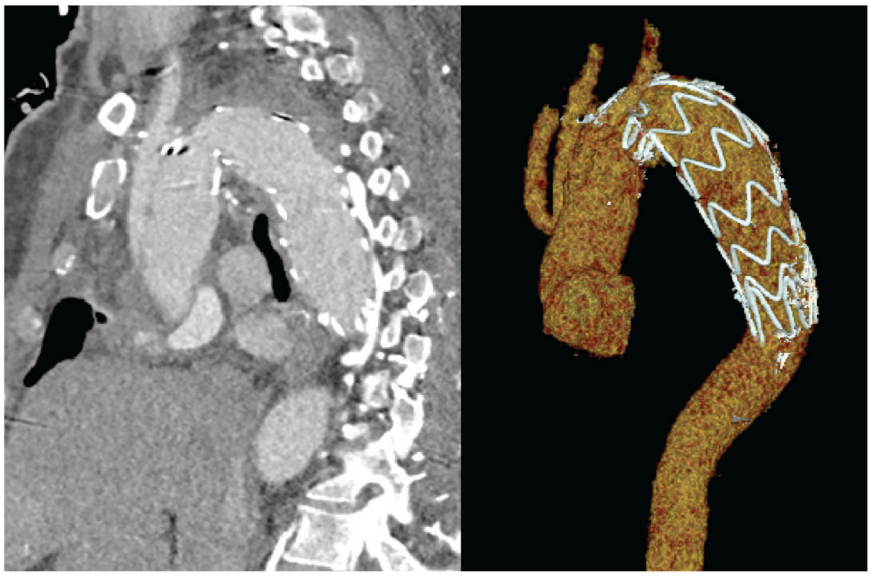

Fig 2. Postoperative computed tomography angiogram of the chest.

be safely feasible, given the proximity of the pseudoaneurysm to the left carotid artery with an inadequate landing zone.[7,8,14]

CONCLUSIONS

A right aortic arch with a retroesophageal left subclavian artery is a rare phenomenon, constituting less than one half of vascular rings. Patients with vascular rings generally undergo repair within the first few years of life; however, aneurysmal aortic changes following the initial repair have been reported.[9] Generally, such cases have been repaired through a right posterolateral thoracotomy.[9] Our experience represents a safe, reproducible repair for a rare and complex pathology.

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