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

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Abstract

A 53 year old male with a history of vascular ring repair secondary to a right sided aortic arch with 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 in the area of his previously divided ligamentum. The patient underwent a successful two-stage repair including a left carotid to subclavian bypass followed by total arch replacement with frozen elephant trunk. He recovered well postoperatively and computed tomography showed complete repair of the pseudoaneurysm with patent bypass graft.

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Abstract

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Introduction

Vascular rings are present in less than 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 a thoracotomy until the early 1990s when endovascular repair techniques emerged[2]. The majority of data in long term follow up discusses persistent tracheal or esophageal compression and concomitant cardiovascular reoperations; however, little data is present for long term vascular complications from arch anomalies and their management[3].

Case Description

A 53 year-old man presented with a large aortic pseudoaneurysm of his distal aortic arch. His past medical history was significant for a right-sided aortic arch with retroesophageal left subclavian artery with a ligamentum arteriosum originating from the descending aorta. He underwent a repair via left thoracotomy at 6 years of age with division of the ligamentum. It was unknown if he had a concomitant Kommerell diverticulum resection at that time. He presented with an acute onset of right-sided chest pain. A CT 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 (Figure 1, Figure 2). 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-subclavian bypass with retrograde embolization of the proximal left subclavian artery. Next we performed a total arch replacement with frozen elephant trunk. We approached his aorta through a midline sternotomy. A moderate amount of hemopericardium was present consistent with his diagnosis. Central cannulation was performed with dual stage venous canulation and aortic cannulation proximal to the first branch, the left common carotid artery. A Glidewire was advanced into the distal arch under TEE guidance from a left common femoral artery sheath. He was cooled to deep hypothermia (18 degrees C). His innominate vein was divided for added exposure.

Once 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 commenced and the ascending aorta and aortic arch were opened. 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 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. 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, Sunrise, Florida) 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 re-establish perfusion down the arm. A post-operative angiogram demonstrated exclusion of the pseudoaneurysm, patent reconstruction, and no evidence of endoleak. His post-operative course was complicated by graft thrombosis of his left common carotid graft which required surgical revision on postoperative day 1, and respiratory failure requiring tracheostomy. 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 endoleak (Figure 3,4).

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]. Vascular rings are formed due to abnormalities in development of the six pairs of aortic arches[5]. When the left fourth arch involutes, a right aortic arch is formed with the apex to the right of the trachea[6]. The two variations are retroesophageal left subclavian artery (65%) and mirror-branching (35%)[7]. The subclavian artery originates from the descending aorta and courses to the left behind the esophagus. Meanwhile, the ligamentum extends from the descending aorta to the left pulmonary artery, completing the vascular ring. Repair of a right aortic arch with left ligamentum and retroesophageal left subclavian artery is via a left thoracotomy in. the fourth intercostal space followed by ligation of the ligamentum arteriosum[6]. Patients with a right aortic arch and left ligamentum may have a Kommerell's diverticulum at the origin of the left subclavian artery from the descending aorta. In these cases, the diverticulum is resected, and the left subclavian artery is transferred to the left carotid artery[1]. In one review of patients with right sided aortic arch and Kommerell's diverticulum, 6% of patients presented with rupture and 47% with dissection[8].

Several operative strategies may be considered depending on clinical presentation including endovascular, hybrid, or open. Resection of the pseudoaneurysm from a right thoracotomy is technically feasible; however, we felt that given the size and location of the pseudoaneurysm that this would be very challenging. Other repair strategies include total arch replacement, descending thoracic aortic replacement with subclavian-carotid transposition, and stent grafting with extra-anatomic bypass, which was performed in this case[9]. The frozen elephant trunk approach allowed for exclusion of the descending pseudoaneurysm and treatment of the head vessels in our patient; however, staging with extraanatomical bypass minimized circulatory arrest time and address the left subclavian aneurysm.

Right aortic arch with retroesophageal left subclavian artery is a rare phenomenon making up less than half of vascular rings. Patient's with vascular rings are generally repaired within the first few years of life; however, aneurysmal aortic changes following initial repair have been reported[8]. Generally such cases have been repaired through a right posterolateral thoracotomy[8]. Our experience represents a safe and durable repair for a rare and complex pathology.

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Author contributions

Aaron J Clark MD – concept, drafting article, critical revision of article.

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Suraj Parulkar MD – critical revision of article,

Christopher K Mehta MD - concept, critical revision of article, approval of article

Figures

1) Preoperative computed tomography sagittal view of pseudoaneurysm

2) Preoperative computed tomography coronal view of pseudoaneurysm

3) Postoperative computed tomography sagittal view of repair

4) Postoperative computed tomography three dimensional reconstruction of repair