Hybrid repair of an adult with a double aortic arch, coarctation of the aorta, and left subclavian artery aneurysm

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ABSTRACT

We report the repair of a double aortic arch, coarctation of the aorta, and left subclavian artery (LSCA) aneurysm using a hybrid procedure in a 47-year-old woman. The patient underwent repair through a median sternotomy incision to bypass the left common carotid artery and LSCA. An endovascular approach was used to repair the coarctation of the aorta and to occlude the right aortic arch. Repair of this anomaly was advised because of worsening clinical symptoms and potential for fatal rupture or dissection of the LSCA aneurysm. Hybrid repair simplified what would have required a multistage open repair. (J Vasc Surg Cases and Innovative Techniques 2019;5:535-7.)

Keywords: Double aortic arch; Hybrid repair; Vascular ring; Coarctation; Aneurysm; Pseudoaneurysm

A double aortic arch, one of the most common forms of vascular ring, occurs when the aortic arch and its branches encircle and compress the trachea, esophagus, or both.[1] The "gold standard" for treatment of these patients was open repair until the early 1990s, when endovascular strategies emerged.

The patient has provided consent to have her case and images published.

CASE REPORT

Our case involves a 47-year-old woman reporting chest pain, headaches, dysphagia, and intermittent hoarseness. Radiographic imaging demonstrated a complete vascular ring with double aortic arch configuration (Figs 1 and 2). There was aortic coarctation at the level of the ligamentum arteriosum and aneurysmal dilation of the proximal left subclavian artery (LSCA) measuring 3.9 × 3.6 cm. The patient's hoarseness was likely secondary to the dilation of the LSCA, compressing the recurrent laryngeal nerve. There was a left dominant aortic arch with a smaller right aortic arch. The right nondominant arch supplied the right vertebral artery (RVA) and passed between the esophagus and the vertebral body (Fig 3). The innominate artery came off the left aortic arch and provided a normal right subclavian artery and right common carotid artery. Preoperatively, transesophageal echocardiography indicated the presence of a bicuspid aortic valve without stenosis or insufficiency. There were no gradients across the coarctation, probably because of the compensating flow provided by the right aortic arch that drained distal to the coarctation. There was discrete narrowing of the left dominant arch. The descending aorta was normal below the coarctation. The abdominal aorta showed a small, 1.9- × 2.6-cm distal saccular aortic pseudoaneurysm (Fig 4). Computed tomography (CT) scan of the head showed no aneurysmal dilation of intracranial vessels. Repair of the double aortic arch, coarctation, and LSCA aneurysm anomaly was advised to relieve symptoms of headache and hypertension and to avoid the risk of rupture or dissection of the aneurysmal LSCA.

Intraoperatively, a spinal fluid drain was placed following institutional protocol, although we no longer place one for stents above the diaphragm. The left common femoral artery was accessed percutaneously and the right common femoral artery was exposed. A supraclavicular incision was used to expose the LSCA aneurysm. The left vertebral artery (LVA) was just distal to the aneurysm. To preserve the LVA, ligature of the LSCA was planned just proximal to the takeoff of this vessel. A primary median sternotomy incision was made, noticing large venous collaterals. The ascending aorta had discrete ectasia without aneurysm. Under a partial occluding clamp, a 14- × 7-mm bifurcated Hemashield graft (Maquet, La Ciotat Cedex, France) was anastomosed from the ascending aorta to the left common carotid artery and LSCA. The left common carotid artery was ligated at the origin and the LSCA was ligated proximal to the LVA. Endovascular closure of the right aortic arch was carried out through the right common femoral artery approach using a 14-mm Amplatzer vascular plug (AGA Medical Corporation, Plymouth, Minn). The plug was delivered to the left of the takeoff of the RVA. The RVA is fed retrograde through the innominate artery, which was confirmed with angiography. Vascular closure of the nondominant arch proximal to the RVA would have led to occlusion of this vessel. We then addressed the aortic coarctation and LSCA aneurysm by endovascular means. A 32- × 100-mm Valiant thoracic endovascular

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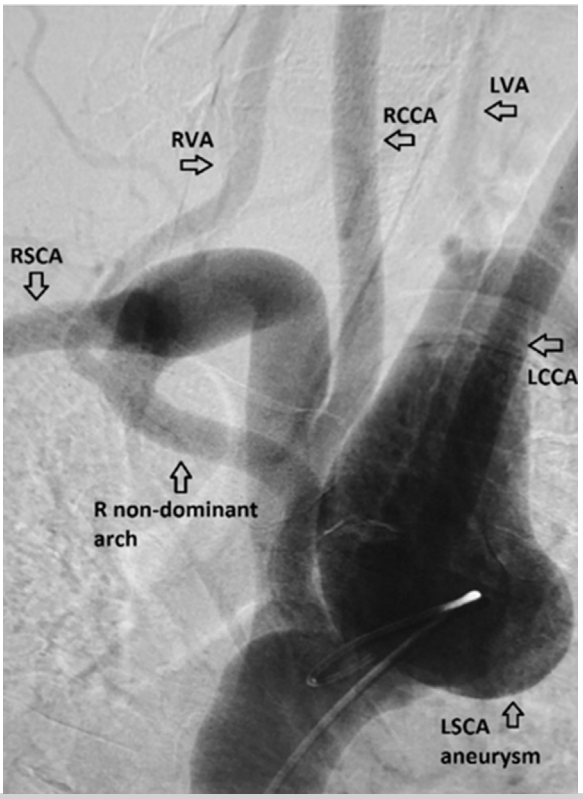

Fig 1. Preoperative angiogram illustrating double aortic arch and aneurysmal dilation of left subclavian artery (*LSCA*). *LCCA,* Left common carotid artery; *LVA,* left vertebral artery; *RCCA,* right common carotid artery; *RSCA,* right subclavian artery; *RVA,* right vertebral artery.

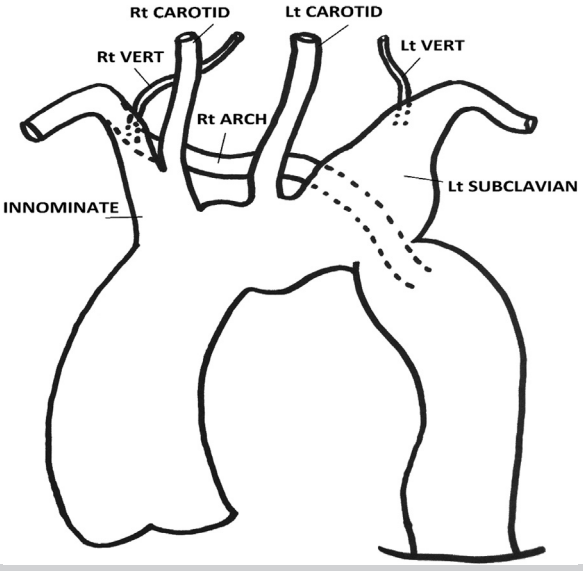

Fig 2. Sketch illustrating preoperative anatomy of the aortic arch.

stent graft (Medtronic, Minneapolis, Minn) was deployed with a proximal landing zone at the level of the innominate artery. The patient had no gradient across the coarctation; therefore, stent

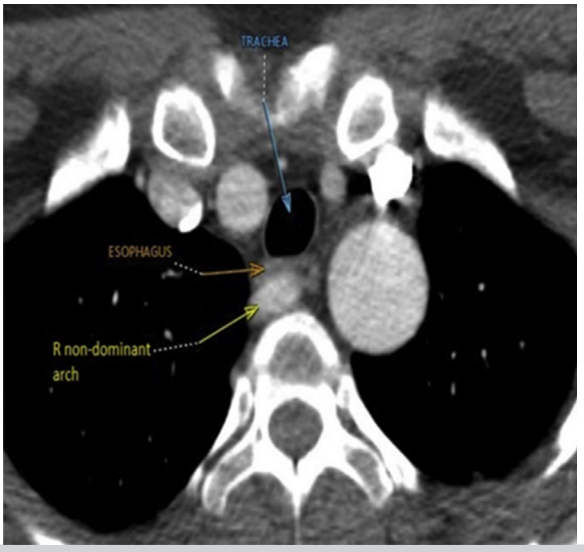

Fig 3. Preoperative computed tomography (CT) scan showing right nondominant arch between esophagus and vertebral body.

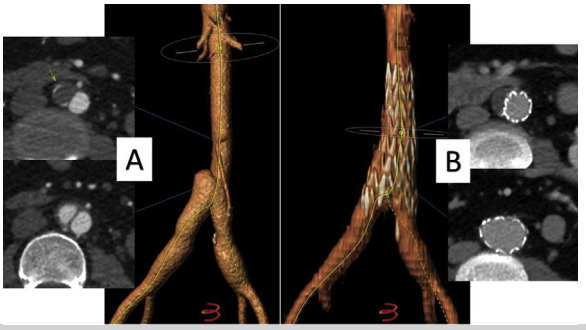

Fig 4. A, Computed tomography (CT) angiogram demonstrating distal abdominal aortic pseudoaneurysm on initial CT (*left upper image*) and at 1-year follow-up (*left lower image*), with three-dimensional reconstruction showing the aneurysm before repair. **B**, After endovascular repair with occlusion of distal abdominal aortic pseudoaneurysm sac.

opposition was completed with a compliant balloon (Reliant; Medtronic) and was confirmed by conventional angiography. Assessment of distal pulses in both upper and lower extremities demonstrated normal pulses with no gradient differences. The patient remained in the intensive care unit for 1 day after surgery and was discharged home by day 5. Her symptoms were resolved by 6 months after the initial intervention. She has remained normotensive, without hoarseness or dysphagia, and no longer has frequent headaches. She was followed up by CT angiography at 4 weeks, at 6 months, and yearly since the repair. The ascending aorta has remained unchanged, and the aortic valve is competent. The LSCA aneurysm is no longer visualized.

A follow-up CT scan performed 1 year after initial repair demonstrated a well-seated stent with complete obliteration of the LSCA aneurysm lumen with no endoleaks (Fig 5). The

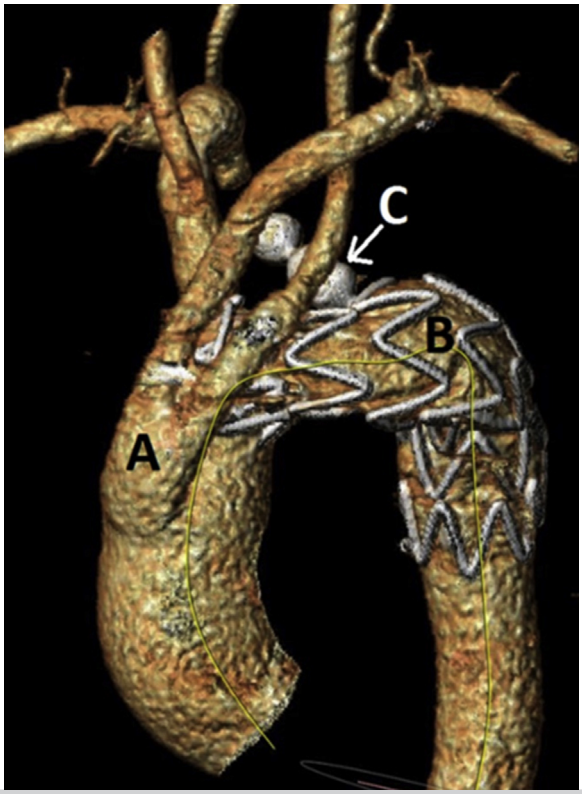

Fig 5. Reconstruction of completion angiogram showing bifurcated graft (*A*), aortic stent (*B*), obliteration of right nondominant arch with Amplatzer device (*C*), and patency of the head vessels.

small saccular pseudoaneurysm first seen in the distal infrarenal aorta showed a new focal dissection with extravasation of contrast material into the sac. An endovascular repair was carried out using a 16- × 28- × 82-mm Medtronic Endurant II stent graft with excellent result (Fig 4).

DISCUSSION

Double aortic arch is rare and is especially unusual in the adult. Compression of the esophagus and trachea can lead to dysphagia, retrosternal pain, cough, and dyspnea. Compression and palsy of the recurrent laryngeal nerve, called Ortner syndrome, leads to hoarseness.[2] Surgical repair of vascular rings is not typically indicated unless the patient is symptomatic or imaging suggests aneurysmal changes.[3] Dysphagia was resolved after obliteration of the nondominant aortic arch. We believe this was due to the lack of blood flow through the posterior arch, relieving the compression between the vertebral body and the esophagus.

Close follow-up of the aortic valve and ascending aorta because of the bicuspid valve disease continues.

We believe debranching through median sternotomy offers a more favorable long-term patency rate in a 47-year-old patient than cervical debranching. Exposure of the ascending aorta would have facilitated repair in the event of bleeding or inability to complete the repair endovascularly. Cervical debranching could have been an option if successful endovascular repair of the right nondominant arch would have been achieved.

Finally, because of the small size of the distal aortic pseudoaneurysm and lack of contrast material within the sac, we thought that early intervention was not needed. However, careful consideration should be given to whether early intervention would be more beneficial. Major variation exists in management of elective abdominal aortic aneurysm (AAA) repair despite established diameter threshold guidelines, with studies demonstrating no significant difference in 1-year mortality for early intervention of small AAAs compared with guideline-size AAAs.[4] Further research correlates these findings, indicating a nonstatistically significant higher survival rate for early intervention of small AAAs compared with surveillance.[5] More aggressive aortic observation in this type of patient could be advised.

CONCLUSIONS

This paper differs from other common vascular anomalies for the rarity of encountering a double aortic arch in an adult, particularly in combination with an aortic coarctation. A hybrid repair seems a better approach than conventional open repair, with fewer comorbidities, including recurrent laryngeal nerve injury, hemorrhage, and need for cardiopulmonary bypass.

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