

Case Report

Open surgical repair of a cervical aortic arch aneurysm: a case report

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Abstract: Upwardly displaced cervical aortic arch (CAA) is an uncommon congenital anomaly. In approximately 20% of cases, it includes an arch aneurysm which is most often situated between the origins of the left common carotid and subclavian arteries. While CAA alone is generally asymptomatic, dysphagia, dyspnea, and recurrent pulmonary infections can occur when an aneurysm is present. We report the surgical repair of a CAA and aneurysm in a 19-year-old male. The left aorta and the descending thoracic aorta were replaced under direct vision without complications, and the patient was discharged on postoperative day 10.

Keywords: Aneurysm, cervical aortic arch, graft

Introduction

A cervical aortic arch (CAA) is generally located between the right brachiocephalic and the left subclavian arteries, with approximately 150 reported cases [1, 2]. A CAA accompanied by an aneurysm is unusual. Herein, we report the surgical repair of a CAA and aneurysm in a 19-year-old male.

Case report

A 19-year-old male was referred for evaluation of a heart murmur detected during a school health examination. He denied any symptoms or medical problems. Our examination revealed a pansystolic III-IV/VI murmur at the left sternal border. His heart rate was 76 beats/min and blood pressure (BP) was approximately 130/80 mmHg in the upper limbs and 136/89 mmHg in the lower limbs. Cardiac color Doppler showed severe narrowing in the aortic isthmus with a tortuous course and dilatation distal to the isthmus. No heart abnormalities were noted (**Figure 1A, 1B**). Computed tomography angiography (CTA) revealed the left common carotid artery (CCA) was crooked and narrow at the thoracic aortic isthmus and at the beginning of the left

subclavian artery and locally expanded. The diameter of the normal aorta was 1.6 cm. Collateral circulation was also observed. Chest X-ray showed that the aortic knob was shortened (**Figure 1C**). In anticipation of surgery, enhanced magnetic resonance imaging (MRI) found the circle of Willis to be complete.

Surgery was performed in the right lateral decubitus position under extracorporeal circulation via cannulation of the right femoral artery. A lateral incision was made at the 4th rib, and the left aortic arch appeared as a grape-like prominent pouch. A clamp was placed between the left CCA and the right brachiocephalic trunk to encompass the stenotic portion of the descending aorta which extended well beyond the aneurysmal neck, leaving enough space for the anastomosis between the graft and the proximal aortic wall. In this manner, the beating heart supplied arterial blood to the brain through the right carotid and vertebral arteries, and the blood supply to the lower organs was via femoral artery catheterization. The proximal aneurysm neck was cut off and a No. 14 MAQUET single-branch vascular graft (~5 mm in diameter) was anastomosed in an end-to-end manner to the left subclavian artery.

Cervical aortic arch aneurysm

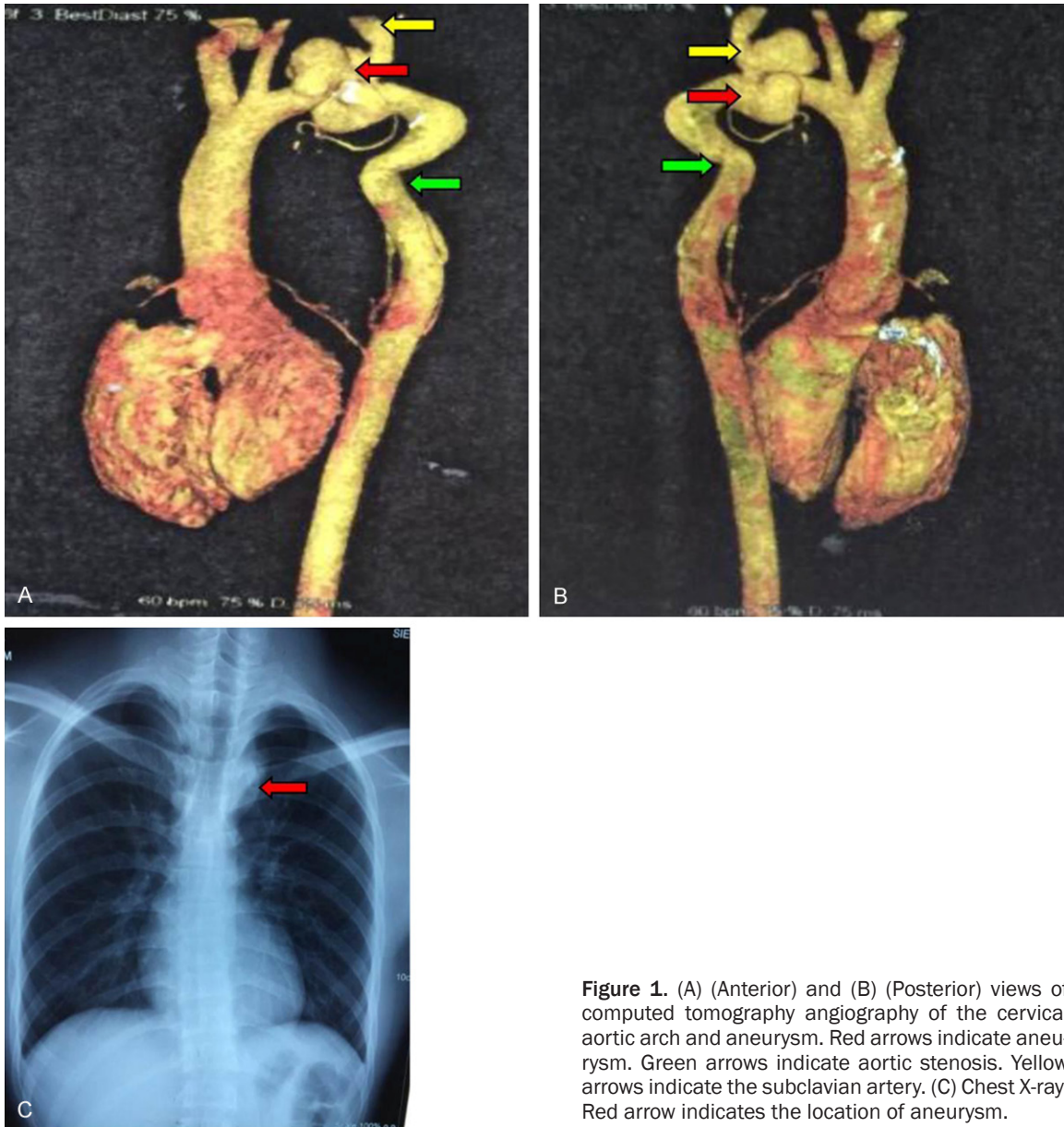


Figure 1. (A) (Anterior) and (B) (Posterior) views of computed tomography angiography of the cervical aortic arch and aneurysm. Red arrows indicate aneurysm. Green arrows indicate aortic stenosis. Yellow arrows indicate the subclavian artery. (C) Chest X-ray. Red arrow indicates the location of aneurysm.

The vascular graft was clamped, and the proximal clamp was opened as soon as the proximal anastomosis was completed. Air in the blood vessel was removed, and the left CCA was opened to restore the blood supply to the brain. The distal aneurysm neck was cut off, and end-to-end anastomosis was carried out between the vascular graft and aorta (Figure 2). Postoperative mean arterial pressure was 85 mmHg in the right upper limb and 78 mmHg in the right lower limb. Microscopic examination of the surgical specimen after hematoxylin-eosin staining revealed disordered arrangement and mucoid degeneration of the smooth

muscle in the aorta wall. His postoperative course was uneventful, and he was discharged 10 days after surgery. Follow-up 3-dimensional spiral CT reconstruction at the time of discharge showed unobstructed blood flow in the graft.

Discussion

CAA is a rare congenital upward displacement of the aortic arch, and aneurysms occur in approximately 20% of cases [3]. Most CAAs are asymptomatic, but dysphagia, dypnea, or recurrent pulmonary infections may occur when an

Cervical aortic arch aneurysm

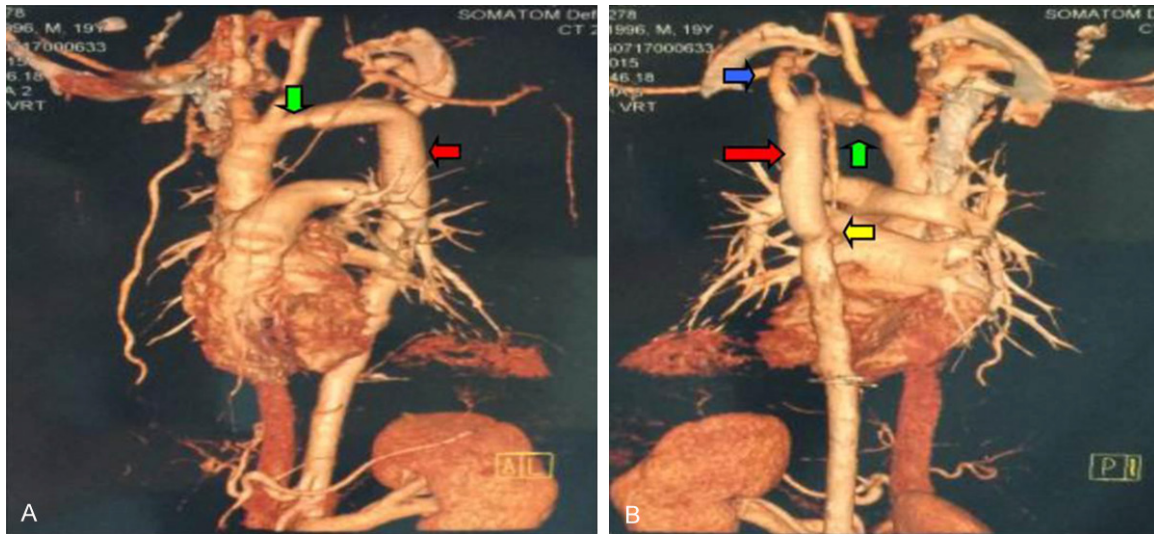


Figure 2. Postoperative computed tomography angiography. (A) Anterior view and (B) posterior view. Red arrow indicates the vascular graft. Green arrow indicates anastomosis between the vascular graft and the left-sided aortic arch. Yellow arrow indicates anastomosis between the vascular graft and the descending aorta. Blue arrow indicates anastomosis between the vascular graft and the left subclavian artery.

aneurysm is present [3]. The sex distribution of CAAs is almost equal, but aneurysms are more common in females [2]. Arterial abnormalities, including congenital coarctation of the aorta, are more common in males [3]. Aneurysms in adults generally develop due to atherosclerosis. In infants, abnormal collagen affects the integrity of the arterial wall structure. Factors associated with the development of CAAs include abnormal embryonic development, connective tissue diseases, hemodynamic changes and arterial wall pressure, and trauma [4]. The 22q11.2 deletion has been associated with congenital cardiac anomalies, including CAA [5], though it was not observed in our case. Cardiac Doppler ultrasound, CTA, and digital subtraction angiography are useful for diagnosis.

Surgical correction can be performed with the patient supine using a midline sternotomy or in the lateral recumbent position. The supine position allows dissection of the aneurysm near the left CCA; however, aneurysm of the thoracic descending aorta cannot be easily dissected. The lateral recumbent position allows dissection of the thoracic descending aorta, but does not allow access to the left CCA. Endograft treatment was not considered in our case because the inner diameter of the aortic arch was small and the course was tortuous,

which prevented passage of a stent delivery system.

During surgery, the vascular clamp on the aorta should be located between the left CCA and the right brachiocephalic trunk to avoid slippage. As much of the aneurysm tissues should be removed to reduce the chance of postoperative recurrence, and the size of the vascular graft should meet that of the normal blood vessel. Although 1.6 cm is somewhat small aortic diameter, this is normal for a 19-year-old Asian youth 168 cm in height. We tend to use relatively small artificial vessels as we have noted that anastomotic bleeding is more common when the grafts are relatively large.

Conclusion

A CAA with aneurysm is a rare condition that can be treated successfully with appropriate preoperative and surgical management.

Acknowledgements

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Disclosure of conflict of interest

None.

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