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**Cervical aortic arch with aneurysm formation and an anomalous right subclavian artery and left vertebral artery: A case report**

Wu YK *et al*. CAA with aneurysm RSA and LVA

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**Abstract**

BACKGROUND

A cervical aortic arch (CAA) refers to a high-riding aortic arch (AA) that often extends above the level of the clavicle. This condition is very rare, with an incidence of less than 1/10000.

CASE SUMMARY

A 29-year-old woman was admitted to the otolaryngology department of our hospital for repeated bilateral purulent nasal discharge for the prior 3 mo. The patient was diagnosed with chronic sinusitis and chronic rhinitis at admission. A preoperative noncontrast chest computed tomography scan showed a high-riding, tortuous AA extending to the mid-upper level of the first thoracic vertebra with local cystic dilatation. A further computed tomography angiography examination showed that the brachiocephalic trunk, left common carotid artery, left vertebral artery (LVA) (slender), and left subclavian artery sequentially branched off of the aorta from the proximal end to the distal end of the AA. The proximal end of the right subclavian artery (RSCA) was tortuous and dilated. The AA showed tumor-like local expansion, with a maximum diameter of approximately 4 cm. After consultation with the department of cardiac macrovascular surgery, the patient was diagnosed with left CAA with aneurysm formation and an anomalous RSCA and LVA and was transferred to that department. The patient underwent AA aneurysm resection and artificial blood vessel replacement under general anesthesia and cardiopulmonary bypass. No abnormality was found during the 2-mo follow-up after discharge.

CONCLUSION

A CAA is a rare congenital anomaly of vascular development. The present unique case of CAA with aneurysm formation and an anomalous RSCA and LVA enriches existing CAA data.

**Key Words:** Cervical aortic arch; Aortic aneurysm; Aortic anomaly; Computed tomography angiography; Haughton classification; Case report

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**Core Tip:** A cervical aortic arch (CAA) is a rare congenital anomaly of vascular development and refers to a high-riding aortic arch that often extends above the level of the clavicle. This paper reports a case of a unique CAA with aneurysm formation and an anomalous right subclavian artery and left vertebral artery. The present unique case enriches existing CAA data.

**INTRODUCTION**

A cervical aortic arch (CAA) is a rare congenital anomaly of vascular development and refers to a high-riding aortic arch (AA) that often extends above the level of the clavicle[1-4]. This paper reports a case of a unique CAA with aneurysm formation and an anomalous right subclavian artery (RSCA) and left vertebral artery (LVA).

**CASE PRESENTATION**

***Chief complaints***

A 29-year-old woman presented repeated bilateral purulent nasal discharge for 3 mo.

***History of present illness***

The symptoms started 3 mo prior to presentation, with dizziness and headache.

***History of past illness***

The patient underwent a cesarean birth at a local hospital 4 years prior.

***Personal and family history***

The patient had no family history that was related to the present illness.

***Physical examination***

Physical examination showed a slightly congested mucosa of the bilateral nasal cavity, a slightly enlarged inferior turbinate, a hypertrophic middle turbinate, and a small amount of purulent nasal discharge in both middle nasal passages and posterior nostrils.

***Laboratory examinations***

Laboratory tests showed that the serum uric acid (428.9 μmol/L), fibrin (4.9 g/L), erythrocyte sedimentation rate (30 mm/h) and ultrasensitive thyroid stimulating hormone (5.1 μIU/mL) values were slightly elevated, the myoglobin (< 21 ng/mL) level was slightly decreased, and the complete blood count, electrolyte profiles and liver function were normal.

***Imaging examinations***

A computed tomography scan at another hospital showed bilateral maxillary sinusitis and ethmoiditis and bilateral inferior turbinate hypertrophy. The patient was diagnosed with chronic sinusitis and chronic rhinitis at admission. A preoperative noncontrast chest computed tomography scan showed a high-riding, tortuous AA extending to the mid-upper level of the first thoracic vertebra with local cystic dilatation, and a few calcified plaques were identified in the descending aorta (DA). A further computed tomography angiography examination showed that the ascending aorta was located to the right of the spine. The brachiocephalic trunk (BCT), left common carotid artery (LCCA), LVA (slender), and left subclavian artery (LSCA) sequentially branched off of the aorta from the proximal end to the distal end of the AA. The BCT bifurcated into the right common carotid artery and the RSCA. The proximal end of the RSCA was tortuous and dilated (approximately 1.5 cm in diameter). The AA showed tumor-like local expansion, with a maximum diameter of approximately 4.0 cm. The long and tortuous AA distal to the aneurysm was located to the left of the spine, extended upward, surpassed the level of the clavicle, and reached the mid-upper level of the first thoracic vertebra. Then, it extended downward tortuously along the left side of the spine to become the DA (Figure 1).

**FINAL DIAGNOSIS**

The final diagnosis of the present case was left CAA with aneurysm formation and an anomalous RSCA and LVA.

**TREATMENT**

After consultation with the department of cardiac macrovascular surgery, the patient was diagnosed with left CAA plus aneurysm formation and an anomalous RSCA and LVA and was transferred to that department. The patient underwent AA aneurysm resection and artificial blood vessel replacement under general anesthesia and cardiopulmonary bypass (CPB) on July 10, 2021. The left CAA and the aneurysm (with a maximum diameter of approximately 4.0 cm) that had formed between the LCCA and the LSCA were observed during the operation. During the intraoperative period, a total of 2 units of type O suspended red blood cells, 10 units of cryoprecipitated antihemophilic factors, and 500 mL of fresh frozen plasma were transfused, and vasopressor drugs, including 8 µg deoxyepinephrine, 128 µg adrenaline, and 38478 µg dopamine, were infused with an infusion pump control to manage blood pressure. The total CPB time was 106 min, the ascending aorta was blocked for 44 min, and circulation was stopped for 15 min. After successful surgery, the patient received intensive care, anti-infection treatment, respiratory and circulatory function maintenance, and microcirculation improvement therapy for 44 h and 50 min.

**OUTCOME AND FOLLOW-UP**

The total hospital stay was 30 d. The patient’s recovery was smooth, and no abnormality was found at the 2-mo follow-up after discharge.

**DISCUSSION**

CAA was first reported by Reid[5] in 1914. CAA is very rare, with an incidence of less than 1/10000[1,3,6], although it is relatively more common in young women[6]. In the 35 cases of CAA reported by Zhong *et al*[6], the average patient age was 34.2 years, and females accounted for 65.7% of the sample.

The etiology of CAA is unclear but is possibly related to abnormal embryonic development of the AA (persistence of the 2nd or 3rd arch or incomplete descension of the normally developed fourth arch, resulting in incomplete entry of the AA into the thoracic cavity)[7-10]. Chromosome 22q11 deletion is evident in some CAA cases[3,4,11].

Most CAA cases are asymptomatic and are usually detected incidentally[4,6,12]. A few cases manifest as a pulsatile neck mass[4,6] or dyspnea and dysphagia due to compression of the trachea and esophagus[6,12,13]. Some CAA cases are combined with congenital cardiovascular anomalies, such as patent ductus arteriosus, tetralogy of Fallot, ventricular septal defect, pulmonary atresia[1,14], and various anatomical variations of the AA branches[2,3,13]. Approximately 20% of CAA cases are complicated with aneurysm formation[1,9,11], which is more common in women[8,9]. The aneurysm may be due to abnormal hemodynamics and arterial wall pressure changes caused by embryonic development, connective tissue anomalies, and a long and tortuous aorta[11,14]. Haughton *et al*[15] classified CAA into five types. Type A: The CAA and DA are contralateral, one side of the common carotid artery is missing, and the independent internal and external carotid arteries directly branch from the AA. Type B: The CAA and DA are contralateral, and two common carotid arteries branch from the AA. Type C: The CAA and DA are contralateral, and a bicarotid trunk appears. Type D: The CAA and DA are ipsilateral, and the sequence of brachiocephalic branching is normal. Type E: This type refers to a right CAA with an ipsilateral DA. Haughton *et al*[15] type D is the most common type of CAA that is combined with aneurysms[3,16], most aneurysms are located in the AA between the LCCA and the LSCA[6,8,16], but a few are located in the DA or simultaneously involve the AA and DA[16]. Shayan *et al*[11] reported eight cases of CAA with aneurysms (type D aneurysm in seven cases and type A aneurysm in one case), including six cases in which the aneurysm was located between the LCCA and the LSCA and two cases in which the aneurysm was located between the LSCA and the DA.

The patient described in this study had a left CAA with an ipsilateral DA. Her long and tortuous AA straddled the left side of the spine, extended upward above the clavicle, and then extended downward tortuously along the left side of the spine. The concurrent AA aneurysm was located between the LCCA and the ostium of the LSCA, and the origins of the two blood vessels were far apart. At the same time, the CAA was combined with the proximal dilatation and tortuosity of the RSCA and the anomalous origin of the LVA, reflecting the uniqueness of her manifestations.

**CONCLUSION**

A CAA is a rare congenital anomaly of vascular development. Some CAA cases are combined with congenital cardiovascular anomalies, and approximately 20% of CAA cases are complicated with aneurysm formation. The present unique case of CAA with aneurysm formation and an anomalous RSCA and LVA enriches existing CAA data.

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**Footnotes**

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**Figure Legends**



**Figure 1 Cervical aortic arch in a 29-year-old female patient.** A: Non-contrast-enhanced chest computed tomography (CT) images (axial view) showing that the top of the aortic arch (AA) was at the mid-superior level of the first thoracic vertebra; B: Contrast-enhanced CT images (axial view) showing the same sign as A; C: Non-contrast-enhanced chest CT images (axial view) showing that the largest plane of the aortic aneurysm was roughly the upper plane of the fifth thoracic vertebrae; D: Contrast-enhanced CT images (axial view) showing the same sign as C; E: Volume reconstruction (VR) (left anterior oblique view) showing that the brachiocephalic trunk, left common carotid artery, and left subclavian artery were sequential branches from the proximal end to the distal end of the AA, that the AA showed tumor-like local expansion, and that the AA distal to the aneurysm and the upper descending aorta (DA) were long and tortuous; F: VR (left posterior oblique view) showing that the right vertebral artery (RVA) originated from the right subclavian artery (RSCA), the left vertebral artery (LVA) originated from the top of the AA, the LVA was thinner than the RVA, and the proximal segment of the RSCA was dilated and tortuous; G: VR (anteroposterior view) showing that the top of the AA was located above the clavicle; H: Curved planar reconstruction showing that the AA straddled the left side of the spine and that the AA distal to the aneurysm and the upper segment of the DA were long and tortuous and were located to the left of the spine.