Right cervical aortic arch with aneurysm and stenotic tract of left descending aorta and aberrant left subclavian artery: case report of a 13-years-old patient

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Abstract

Background: Cervical aortic arch (CAA) is a rare anomaly that could be associated with aortic stenosis, aneurysm or heart malformations. To correct this anomaly, symptomatic patients undergo surgery, usually consisting of a prosthetic graft repair. Moreover, circulatory arrest and deep hypothermia are often needed. **Case presentation:** A 13-years-old patient underwent correction of an aortic arch stenosis with a post-stenotic aneurysm between the origin of the right carotid artery (RCA) and right subclavian artery (RSA) in a right CAA. A resection with direct end-to-end anastomosis was performed, with mild hypothermic cardiopulmonary by-pass. **Conclusions:** Surgical correction of cervical aortic arch anomalies without the use of prosthetic grafts and circulatory arrest may be a safe alternative approach, especially in the pediatric population.

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ABSTRACT:

Background: Cervical aortic arch (CAA) is a rare anomaly that could be associated with aortic stenosis, aneurysm or heart malformations. To correct this anomaly, symptomatic patients undergo surgery, usually consisting of a prosthetic graft repair. Moreover, circulatory arrest and deep hypothermia are often needed.

Case presentation: A 13-years-old patient underwent correction of an aortic arch stenosis with a post-stenotic aneurysm between the origin of the right carotid artery (RCA) and right subclavian artery (RSA) in a right CAA. A resection with direct end-to-end anastomosis was performed, with mild hypothermic cardiopulmonary by-pass.

Conclusions: Surgical correction of cervical aortic arch anomalies without the use of prosthetic grafts and circulatory arrest may be a safe alternative approach, especially in the pediatric population.

KEY WORDS: cervical aortic arch; aneurysm; pediatric aortic arch anomaly.

Introduction

Cervical aortic arch (CAA) is a rare anomaly characterized by a high-lying aortic arch extending at or above the level of the medial ends of the clavicles. Anomalies of arch sidedness and branching, as well as structural abnormalities of the aorta, such as stenotic tract, can be present. CAA is usually associated with other malformations of the left ventricle (LV) outflow tract, but it can also appear as an isolated malformation. Here, we present the case of a 13-year-old female with a right CAA with a stenosis distal to the right carotid artery (RCA), a post-stenotic aneurysm and aberrant left subclavian artery (LSA).

Case report

A 13-year-old female, with a history of endoscopic steroid infiltration for a laryngeal angioma, was referred to our hospital in good clinical condition because of a systolic murmur and a thrill on the right side of the neck, near the clavicle. No pressure gradient between the upper and lower body, as well as between her right and left sides, was found at the medical examination but the echocardiography revealed an aortic diastolic run-off. The CT scan showed a right-sided CAA with contralateral descending aorta and aberrant LSA. A stenotic tract (0.4 cm diameter) with a post-stenotic aneurysm forming a posterior loop was evidenced between the origin of the RCA and the right subclavian artery (RSA). Immediately downstream the aneurysm, up to the origin of the aberrant LSA, the aortic diameter resulted evenly reduced (10x11 mm) for about 5 cm (Figure 1 - 2). The patient underwent cardiac catheterization and a gradient of 90 mmHg between the ascending and descending aorta was found. Thus, the patient was referred to surgery.

In median sternotomy, the arch vessels were isolated, and the descending thoracic aorta mobilized down to the origin of the LSA. The aneurysm at the top of the aortic arch appeared thin and transparent walled. A pressure monitoring line was placed directly in the RCA to check the brain perfusion pressure during the Cardio-Pulmonary By-pass (CPB) time.

This was established with a single atrial cannula for venous drainage while arterial perfusion was accomplished through a double cannulation of the ascending aorta and the right common femoral artery. The patient was cooled at a nasopharyngeal temperature of 26°C. The aorta was cross-clamped after the origin of the RCA and after the origin of the RSA, which was separately occluded (Figure 3). The aneurysmatic tissue and the stenotic tract were removed, and a direct end-to-end anastomosis was performed between the ascending and the descending aorta with a running 5-0 polydioxanone suture. After the procedure, no pressure gradient was observed between the ascending and the femoral artery. The patient's post-operative course was uneventful, and she was discharged after 7 days. Histological evaluation of the resected aorta showed diffused and marked fibrosis of the media. The intima was mostly intact with very mild focal hyperplasia and microcalcification. These anatomical findings couldn't be accurately classified.

Discussion

CAA is a rare anomaly which can be found in 1:10,000 live births (1). Reid et al (2) first described it in 1914,

and subsequently Haughton et al (3) and Zhong et al (4) classified it according to the side of the arch and the branching of the head vessels. The case we reported is similar to a type B1-B2 in Zhong's classification (4): right sided CAA, aberrant LSA and a contralateral descending aorta (which should be ipsilateral).

The embryogenesis of this anomaly is unclear; it is probably due to an abnormal resorption of the third aortic arch and involution of the fourth. Alternatively, it could be caused by a failure of migration of the fourth arch from the cervical to the thoracic area (5).

Clinical presentation is heterogeneous, from an incidental finding to a tracheal or esophageal compression. A pulsatile mass in the neck or a systolic murmur, as in our case, may be present. Common signs include hypertensive crisis and discrepancy of blood pressure between the upper and lower extremities. In our case, these signs were not evidenced at the clinical investigation because both the subclavian arteries arose after the stenotic tract. This makes diagnosis quite difficult and only cardiac catheterization disclosed the real clinical situation.

Anomalies of the arch should be investigated also in asymptomatic patients with history of angiomas as our patient, because of their association in some genetic disorders as for instance PHACE syndrome (6).

Various surgical approaches have been proposed but there is no consensus on the most effective one. In some centers a thoracotomy is preferred, a second alternative includes a median sternotomy with possible extension to the bilateral supraclavicular regions or the clamshell incision (2). Because the very high-lying aortic arch, we preferred to perform a median sternotomy to guarantee an ease access to all the aortic branches and to institute the CPB that was crucial to facilitate the aneurysm isolation and to protect the spinal cord during cross clamping. This was accomplished with femoral artery perfusion and systemic mild hypothermia, that are considered the best way to avoid ischemic organ damage.

In most cases, the aortic arch anomalies in these patients are treated with a graft interposition (5) or patch aortoplasty (6), also in pediatric patients. Our case presented enough aortic tissue to perform a direct end-to-end anastomosis. This strategy was considered the best option in a pediatric patient in order to promote the growth of the native aorta and to prevent the development of pseudoaneurysms, that occur in almost 42% of patients treated with patch aortoplasty (7), or mismatch between the prosthetic graft and the native tissues.

In conclusion, CAA requests a customized approach for the heterogeneity of the anomalies. We reported our experience with a case where only native tissue was used for aortic repair. This strategy represents the best option, if the anatomy is favorable, especially in the pediatric population. Lower body perfusion and systemic mild hypothermia should be considered in every case to facilitate surgical isolation of the aneurysm and to prevent ischemic organs and spine damages.

CONFLICT OF INTERESTS

The authors declare that there are no conflicts of interests.

FIGURES

Figure 3 Schematic representation of the clamping site. RSA was separately occluded using a vessel loop.

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