Abnormally proximal aortic origin of the brachiocephalic artery: Surgical implications

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ABSTRACT

Abnormal proximal aortic origin of the brachiocephalic artery is a very rare condition. It can occur in isolation or associated with complex congenital heart disease affecting the right ventricular outflow tract. Its recognition carries relevant surgical implications for the safe conduct of cardiopulmonary bypass and for any surgical procedures that directly involve the proximal ascending aorta and its branches.

Keywords: Ascending aorta anomalies, brachiocephalic artery, cardiopulmonary bypass, embryology of aorta

INTRODUCTION

A myriad of aortic arch variants has been described in different species and their embryological basis has been a source of debate through the years. Anomalies of the aortic arch and great vessels account for one-third of all congenital cardiovascular defects. Arch variants can occur in isolation or as a part of a spectrum in complex congenital heart disease as in some patients with tetralogy of Fallot (TOF).[1] Arch variants occur in about 80% of all cases of 22q11.2 deletion syndrome, suggesting that gene abnormalities are important risk factors for aortic arch development and anomalies. The increasing utilization of multimodal imaging has enabled the identification of these anomalies and facilitated their preoperative planning. Herein, we describe an abnormally proximal origin of the brachiocephalic artery (BCA), highlighting its clinical significance for surgical procedures.

CASE REPORT

We report a case of TOF with a left aortic arch, wherein preoperative cardiac computerized tomography (CT scan) demonstrated an abnormally proximal origin of the BCA within the pericardial cavity, resulting in BCA elongation. This anatomical variation was an incidental finding and did not cause any signs or symptoms. An intraoperative photograph and CT scan with three-dimensional reconstruction are shown in Figure 1. A short distance beyond the origin of the BCA, the walls of the ascending aorta and the BCA were adherent to each other but without luminal communication, which is evident on the CT scan. We could not detect any other abnormalities in the aortic arch system or in the parietal wall thickness of aortic arch arteries. The patient underwent a successful surgical procedure for the correction of the TOF.

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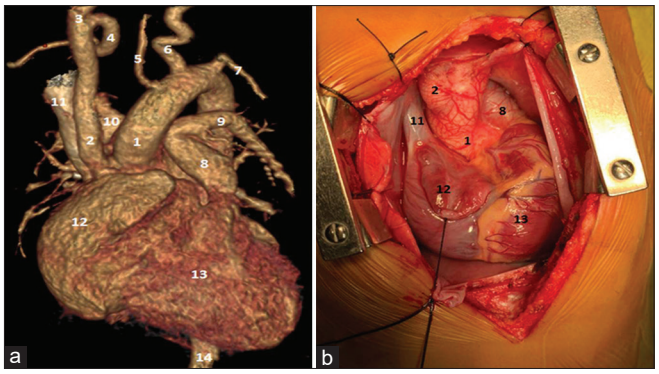

Figure 1: Computerized tomography (CT) scan with three-dimensional reconstruction (a) and intraoperative photograph (b) demonstrating the abnormally proximal aortic origin of brachiocephalic artery (BCA). 1: Ascending aorta, 2: BCA, 3: Right common carotid artery, 4: Right subclavian artery, 5: Left vertebral artery, 6: Left common carotid artery, 7: Left subclavian artery, 8: Main pulmonary artery, 9: Left pulmonary artery, 10: Right pulmonary artery, 11: Superior vena cava, 12: Right atrium, 13: Right ventricle, 14: Descending thoracic aorta. At short distance after the origin of the BCA, the walls of the vessels were adherent to each other without luminal communication, which is evident on CT scan

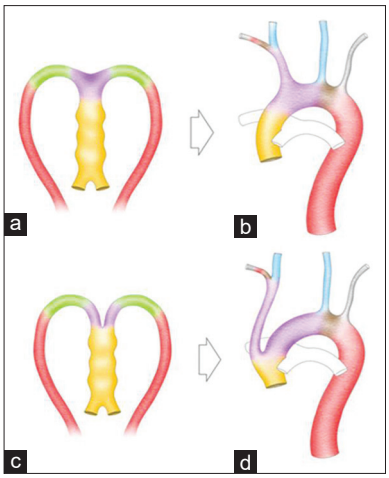

Figure 2: (a) The endocardial tube (in yellow) and the aortic sac (in purple) arise from a single, unpaired vascular plexus that connects with the paired dorsal aortas (in red) via the formation of paired and symmetric third, fourth, and sixth pharyngeal arch arteries (schematically in green). The intrapericardial components of the arterial trunks (ascending aorta and pulmonary trunk) are formed from the outflow tract (yellow), and the aortic sac (purple), on the distal part, contributing to the distal ascending aorta, through a spiraliform septation process. (b) The aortic sac (purple), left horn (purple), and the left fourth pharyngeal arch artery (brown) together form the aortic arch. The right horn (purple) forms the brachiocephalic artery (BCA). As the right-sided third and fourth pharyngeal arch arteries arise from the right horn, the right common carotid and subclavian arteries become the branches of the BCA. (c) In our case, the abnormally proximal aortic origin of BCA can be ascribed to abnormal development of the aortic sac, with a short or absent aortic sac component. (d) During further development, the right horn (purple) forms the BCA with a hyperelongation process, and the left horn (purple) continues as the aortic arch

DISCUSSION

The embryological processes from the aortic arch system development to adult aortic arch transformation, as well as the embryological abnormalities that can lead to congenital cardiovascular defects are complex and difficult to understand. Through the years, many interpretations have been proposed on the basis of inferences from different studies, including various animal models and with the application of different and incomplete techniques.

As far as we know, our case is the first to describe an anomalous origin of the BCA from the proximal portion of the ascending aorta. There has been only a case with an anomalous origin of the common carotid arteries very close to the sinotubular junction,[2] in which the authors suggested a developmental anomaly affecting the aortic sac, with septation of the aortic sac starting in proximity to the orifice of the third, fourth, and sixth aortic arch arteries, thus, accounting for the proximity of the two arteries and the aorta.

The embryological process of the development of the aortic arch system is shown in Figures 2a-d.[3,4]

In our case, a developmental anomaly related to the aortic sac could have caused a lack of tissue in the ascending aorta, accounting for the proximity of the sinotubular junction and the BCA. The lack of tissue could be due to inhibition or absence of the proliferative stimulus and/or hypoplasia of aortic sac cells. It is beyond the scope of the present paper to further discuss and dissect the molecular mechanisms, which determine the aortic arch system formation. However, it can be stated that complex cellular and molecular mechanisms play a crucial role in this remodeling process, and the perturbation of this perfect equilibrium can generate a wide range of abnormalities.

If the BCA arises proximally from ascending aorta as in this case, we would suggest monitoring pressures via a right radial arterial line and a femoral arterial line. After establishing cardiopulmonary bypass (CPB) with the aortic inflow cannula, one can place an additional inflow cannula in the BCA. This is done if lower perfusion pressure in the radial arterial line is noted during cross-clamp. We did not electively choose to cannulate both the aorta and the BCA in the first place. We did not experience any problems with the proximity of the BCA origin to the sinotubular junction in cross-clamping. Although this anatomical aberration, when isolated, does not cause symptoms or clinical issues, prior knowledge helps in planning a safe CPB strategy for the repair of associated cardiac malformations.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Prabhu S, Kasturi S, Mehra S, Tiwari R, Joshi A, John C, *et al.* The aortic arch in tetralogy of Fallot: Types of branching and clinical implications. Cardiol Young 2020;30:1144-50.
2. Boerman RH, Padberg GW, Wondergem J, Gittenberger de Groot AC. Anomalous origin of the common carotid arteries as a cause of stroke. Clin Neurol Neurosurg 1991;93:317-9.
3. DeRuiter MC, Poelmann RE, VanderPlas-de Vries I, Mentink MM, Gittenberger-de Groot AC. The development of the myocardium and endocardium in mouse embryos. Fusion of two heart tubes? Anat Embryol (Berl) 1992;185:461-73.
4. Rana MS, Sizaro A, Christoffels VM, Moorman AF. Development of the human aortic arch system captured in an interactive three-dimensional reference model. Am J Med Genet Part A 2014;164A: 1372-83.