Anomalous origin of the left brachiocephalic artery in the right aortic arch: Is there a method to the madness?

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ABSTRACT

The anomalous origin of the left brachiocephalic artery in a right sided aortic arch is a rare vascular ring which might lead to esophageal compression. The exact embryological origin of this anomaly is still widely debated. We present an infant who presented with esophageal compression symptoms and review the various hypotheses about the embryological origin of this anomaly.

Keywords: Anomalous innominate artery, right aortic arch, vascular ring

INTRODUCTION

Vascular rings are rare congenital anomalies of the aortic arch. The most common forms of complete vascular rings are the double aortic arch[1] and the right aortic arch with aberrant left subclavian artery associated with the left ductus arteriosus or ligamentum arteriosum. They present with tracheal or esophageal compression symptoms or can be asymptomatic. Other vascular rings have been described with much lower incidence. We report a rare variant of the right aortic arch and an aberrant left brachiocephalic artery in an infant and discuss theories of causation.

CASE DETAILS

An 8-month-old female infant with a history of recurrent vomiting and regurgitation of feeds attributed to gastroesophageal reflux was referred for cardiac evaluation of a systolic murmur as well as fast breathing noticed particularly during feeding. Her weight gain was appropriate for age with no other concurrent illnesses.

She did not exhibit any pallor or cyanosis, and all her peripheral pulses were present and equal. She was mildly tachypneic (respiratory rate 50/min) with minimal subcostal and suprasternal retractions but no stridor. On cardiac evaluation, her first heart sound was normal with a wide and fixed splitting of the second heart sound and a grade 2/6 - ejection systolic murmur at the left second intercostal space.

Her echocardiogram revealed a large secundum atrial septal defect with moderate dilatation of the right heart structures. The intracardiac anatomy was otherwise normal. There was a suspicion of the double aortic arch, and hence, a cardiac computed tomogram was obtained to delineate the arch anatomy. This demonstrated a right-sided aortic arch with the first branch being the right common carotid artery, followed by the right subclavian artery. The left brachiocephalic artery arose from the proximal descending aorta and tracked a retroesophageal course before dividing into the left subclavian and common carotid arteries [Figure 1]. The left common carotid artery spanned a length of 9 mm before bifurcation. It measured 4.1 mm proximally before constricting to 1.7 mm just before the bifurcation [Figure 2]. This implied a high possibility of Kommerell's diverticulum with the left ligamentum and

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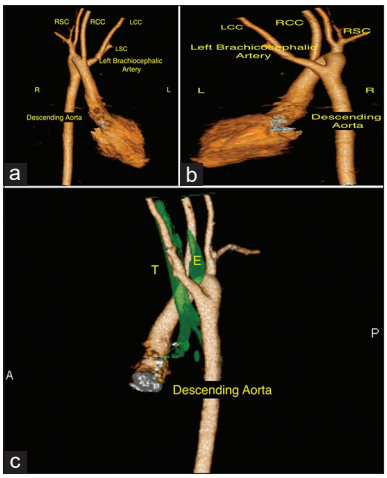

Figure 1: (a and b) Three dimensional reconstruction of computed tomogram demonstrating the aortic arch and branching pattern from the anterior (a) and posterior (b) views. (c) Three dimensional reconstruction demonstrating the retroesophageal course of the left brachiocephalic artery. RCC: Right common carotid, RSC: Right subclavian, LCC: Left common carotid, LSC: Left subclavian, T: Trachea, E: Esophagus

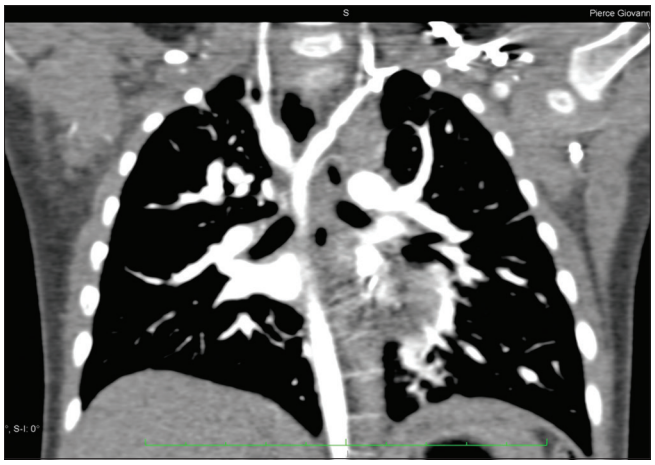

Figure 2: Coronal topogram images demonstrating a constriction in the aberrant left brachiocephalic artery raising the suggestion of a Kommerell's diverticulum

given her esophageal compression symptoms; the patient underwent surgery through a midline sternotomy. A left-sided ligamentum arteriosum was identified and divided during the surgery with concomitant closure of the atrial septal defect.

The postoperative recovery was uneventful, and at follow-up 8 months after surgery, she was asymptomatic with marked improvement in her gastroesophageal reflux.

DISCUSSION

The embryology of this rare vascular ring is best described based on the hypothetical double aortic arch diagram of Edwards.[2] There is presumably regression of the left side of the primitive aortic sac, and the left 4th aortic arch proximal to the 3rd aortic arch resulting in the migration of the left 3rd arch and the left 7th intercostal artery to the left-sided dorsal aorta. A left-sided ligamentum arteriosum from the base of the brachiocephalic artery to the left pulmonary artery completes the ring. Recently, this hypothesis has been challenged. Raimondi *et al.*,[3] suggested that the entire left-sided structures including the left 3rd arch, the left 4th arch as well as the left portion of the truncoaortic sac are absent and that the left head and neck vessels arise directly from the left dorsal aorta without a brachiocephalic artery. They based their hypothesis on the early bifurcation of the left common carotid artery noted in some of their cases. They derived support for their hypothesis from mouse models deficient in homeobox A3 that resulted in failure of formation of the 3rd aortic arch. However, we present a case with a long arterial segment before bifurcation. We speculate that the hypothesis put forward by Raimondi *et al.* does not explain all cases with this anomaly. To the best of our knowledge, there have been no published animal models that elucidate the mechanisms underlying this developmental anomaly of the aortic arch or recapitulate this defect. Our case and other similar reports suggest the possibility that multiple mechanisms underlie the various morphological variations of this anomaly.

We identified <20 reported cases of this anomaly in the literature,[4,5] and the majority of the symptomatic patients described had a typical presentation in adolescence with dysphagia and respiratory symptoms. The infantile presentation of this patient with esophageal symptoms makes it unique among this rare subset. More than half of the patients were asymptomatic and were detected during the evaluation of other congenital cardiac problems.

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Conflicts of interest

There are no conflicts of interest.

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