Left cervical and circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum: A rare vascular ring

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ABSTRACT

We report a rare case of vascular ring due to a left cervical circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum. This is a very rare case of vascular ring in a newborn with stridor and stenosis of the left pulmonary artery. A cardiac catheterization and an angio-computed tomography scan were helpful to clarify the diagnosis. The surgical correction was performed with division of the arterial duct, reimplantation of the right subclavian artery, aortic and tracheal suspension, and pulmonary arterial plasty. The patient was asymptomatic at discharge and 1-year follow-up.

Keywords: Aberrant subclavian artery, cervical aortic arch, circumflex aorta, Kommerell diverticulum, vascular ring

INTRODUCTION

A complete vascular ring is a rare congenital defect in which the trachea and the esophagus are encircled and compressed by anomalous vascular structures (aorta, ductus/ligamentum arteriosus, and aberrant subclavian artery). The two most common types of complete vascular rings are the double aortic arch and the right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum (together 85%–95% of cases).[1] Some patients develop stridor and/or dysphagia due to a direct vascular compression. However, tracheomalacia and bronchomalacia are possible complications related to compression.[2]

We describe a rare case of complete vascular ring due to a left cervical and circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum.

CASE REPORT

A newborn (age 22 days, weight 3.70 kg) came to our attention because of the onset of stridor while crying. Clinical examination did not highlight any significant findings. Echocardiography showed a significant stenosis at the origin of the left pulmonary artery, whereas the aortic arch was never adequately profiled. Cardiac catheterization delineated a very rare vascular ring: A left cervical aortic arch with a right contralateral descending aorta (i.e., circumflex aorta) and an aberrant right subclavian artery arising from a Kommerell diverticulum. A right arterial duct closed the complete vascular ring. The common carotid arteries arose apart from the aortic arch. The pulmonary angiography showed a severe hypoplasia of the left pulmonary branch proximal tract (2 mm, z-score – 5.00) and a mild hypoplasia

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of the distal one (3 mm, z-score – 2.50) [Figure 1]. The angio-computed tomography scan highlighted a direct compression of the right bronchus (deployed between the right pulmonary artery, the arterial duct, the Kommerell diverticulum, and the right subclavian artery) and of the esophagus and trachea (due to the descending aorta crossing the midline posteriorly to the tracheal carina). The stenosis of the left pulmonary artery did not seem to be originated by a compression of the adjacent structures [Figure 2].

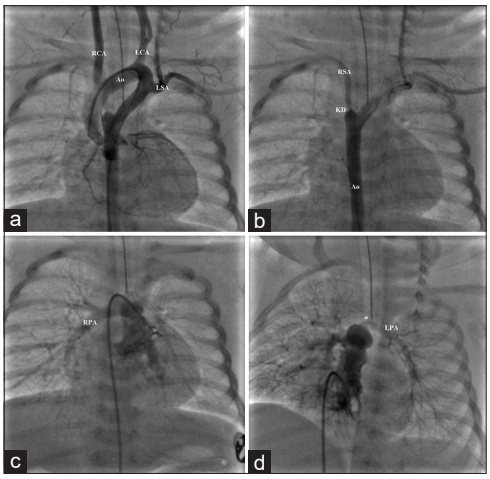

Figure 1: Ascending (a) and descending (b) aortography in anteroposterior view showed: a cervical left aortic arch (overriding the left bronchus) with right-sided descending aorta (circumflex aorta), with an aberrant right subclavian artery arising from a Kommerell diverticulum. A right arterial duct was recanalized with a multipurpose catheter from the Kommerell diverticulum (c). Right ventriculography (d) showed an angulated origin of the left pulmonary artery with significant hypoplasia. Ao: Aorta, KD: Kommerell diverticulum, LCA: Left common carotid artery, LPA: Left pulmonary artery, LSA: Left subclavian artery, RPA: Right pulmonary artery, RSA: Right subclavian artery, RCA: Right common carotid artery

The surgical correction was performed on the day of life 30 with a median sternotomy and without cardiopulmonary bypass.[3] The arterial duct was ligated and divided. The right subclavian artery was divided and implanted to the right common carotid artery with an end-to-side anastomosis. The proximal hypoplastic tract of the left pulmonary artery was enlarged with an autologous pericardial patch. Finally, both an aortic (aortopexy) and a tracheal (tracheopexy) suspension were performed to reduce the compression on the trachea due to the circumflex aorta. Both the aorta and the trachea were suspended to the posterior surface of the sternum with a prolene 6/0 suture. The patient had a normal postoperative course and was discharged 7 days after the surgical correction, without any symptoms. At 1-year follow-up, the patient was asymptomatic, and the echocardiography showed a normal left pulmonary artery and a patent right subclavian anastomosis.

DISCUSSION

Vascular rings in the setting of left aortic arch are rare. Berman *et al.*[4] and Whitman *et al.*[5] described the first cases of complete vascular ring due to a left circumflex aortic arch. Our report is a very rare case of cervical left aortic arch with right-sided descending (circumflex) aorta, aberrant right subclavian artery arising from a Kommerell

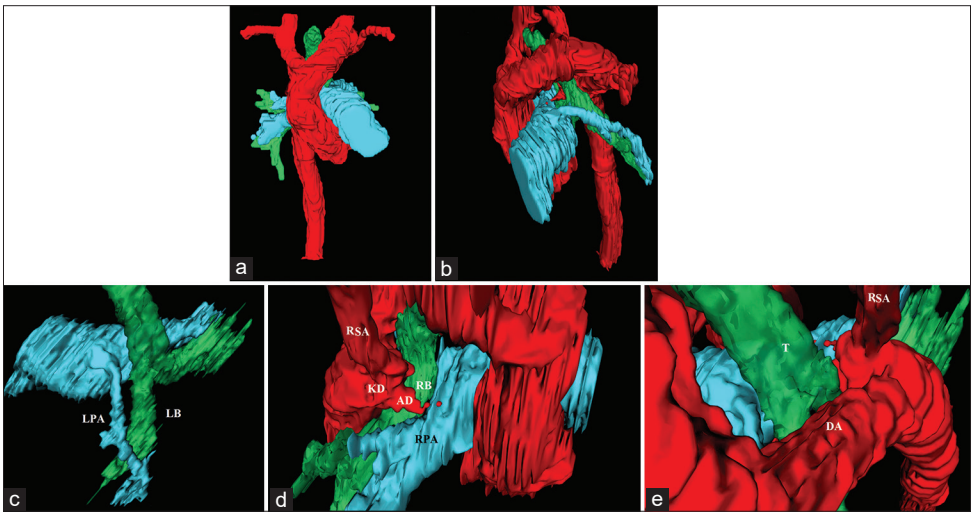

Figure 2: Three-dimensional angio- CT scan in frontal (a) and left anterior oblique (b) views confirmed a complete vascular ring composed by a left cervical and circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum. Detailed window showed: the absence of direct compression between the left bronchus and the left pulmonary artery (c), a direct compression of the right bronchus by the vascular ring (Kommerell diverticulum, arterial duct, and right pulmonary artery) (d), and a direct compression of the trachea by the circumflex descending aorta crossing posteriorly (e). AD: Arterial duct (ligamentum arteriosum), DA: Descending aorta, KD: Kommerell diverticulum, LB: Left bronchus, LPA: Left pulmonary artery, RB: Right bronchus, RPA: Right pulmonary artery, RSA: Right subclavian artery, T: Trachea

diverticulum, and right arterial duct. Zhong *et al.*[6] and Haughton *et al.*[7] identified this vascular anomaly as type B4 (left-sided arch with contralateral descending aorta) and type B (contralateral descending aorta and dual common carotid arteries) cervical aortic arch, respectively. This aortic arch anomaly is extremely rare, and only a few reports are mentioned in the literature.[5,8] The complete vascular ring composed by the right pulmonary artery, right arterial duct, and Kommerell diverticulum realized a direct compression to the right main bronchus. The anomalous course of the descending aorta (crossing the median line behind the carina) determined a posterior compression of the esophagus and trachea. The arterial was ligated and divided in order to release the main right bronchus. Aortopexy and tracheopexy were performed to remove the esophageal and tracheal compression due to the circumflex descending aorta. The surgical "aortic uncrossing" was taken into consideration.[9] However, a less demolitive surgical approach (tracheal and aortic suspension) was preferred as first-line treatment. The aortic uncrossing was considered as an alternative in case of failure of the previous surgical strategy. The right subclavian artery was implanted to the right common carotid artery restoring an iatrogenic "right brachiocephalic artery." Since the stenosis of the pulmonary artery was significant, a surgical pulmonary arterioplasty was performed with a pericardial patch.

CONCLUSIONS

This is an unusual and complex case of complete vascular ring associated with significant stenosis of the left pulmonary artery in a newborn with stridor. The vascular ring was constituted by a left cervical and circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum and a right ductus arteriosus. The surgical correction was performed with the division of the arterial duct, the reimplantation of the right subclavian artery, an aortopexy and tracheopexy, and a pulmonary arterioplasty.

Informed consent

Informed consent for publication as a case report was obtained from the family.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the family has given consent for images and other clinical information to be reported in the journal. The family understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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