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ANATOMIC VARIATIONS

Crossed pulmonary arteries as additional cause of dysphagia in association with right aortic arch and Kommerell diverticulum

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Abstract

We describe an uncommon association of crossed pulmonary arteries and a right aortic arch with a Kommerell diverticulum and a left ligamentum arteriosum, resulting in disabling dysphagia in a 33-year-old woman. First, endovascular exclusion of the Kommerell diverticulum was performed using a thoracic stent graft, associated with left subclavian–carotid transposition. Second, open aneurysmorrhaphy and division of the left ligamentum arteriosum allowed a proper release of the oesophageal compression. Dysphagia completely disappeared in the postoperative course. Control computed tomography angiography at 6-month follow-up showed a satisfactory hybrid repair. A complete understanding of the combined effects of these two anatomical variations on oesophageal compression led to a suitable surgical management.

Keywords Right aortic arch · Kommerell diverticulum · Dysphagia · Left ligamentum arteriosum, vascular ring

Introduction

Crossed pulmonary arteries (CPA) are a rare form of congenital anomaly where both pulmonary arteries cross each other before reaching their respective pulmonary hilum. This uncommon anatomic variation has been reported mostly in newborns and children, in association with cardiac malformations such as persistent truncus arteriosus, and tetralogy of Fallot and DiGeorge syndrome. Nevertheless, reports in the literature are few, especially in adults. We describe a rare case of CPA in a 33-year-old woman who presented with a symptomatic Kommerell diverticulum in a right aortic arch. To the best of our knowledge, this anatomic association has never been reported so far. Furthermore, CPA might play an additional role in the pathophysiology of dysphagia when associated with such an anatomic configuration of the aortic arch.

Case report

A 33-year-old female patient was referred to our institution for severe dysphagia related to a Kommerell diverticulum. Dysphagia onset was progressive and began 2 years before referral, with an associated 7-kg weight loss over the past 6 months. Esogastric endoscopy showed no evidence of oesophageal disease. Computed tomography angiography (CTA) showed a 25-mm Kommerell diverticulum at the level of an aberrant left subclavian artery (LSA) in a right aortic arch. The supra-aortic trunks originated in the following order: the left common carotid artery (LCCA), the right common carotid artery, the right subclavian artery, and the aberrant LSA. In addition, CTA revealed crossed pulmonary arteries (Video—Electronic supplementary material). The bifurcation of the pulmonary trunk lies in a sagittal plane. The left pulmonary artery originated from the right part of the pulmonary trunk, superior to the origin of the right pulmonary artery. The right pulmonary artery coursed from the left and inferior aspects of the left pulmonary artery. Transthoracic echocardiogram revealed no other cardiac or vascular associated malformation. As shown in Fig. 1, the oesophagus was severely compressed, presumably by a left

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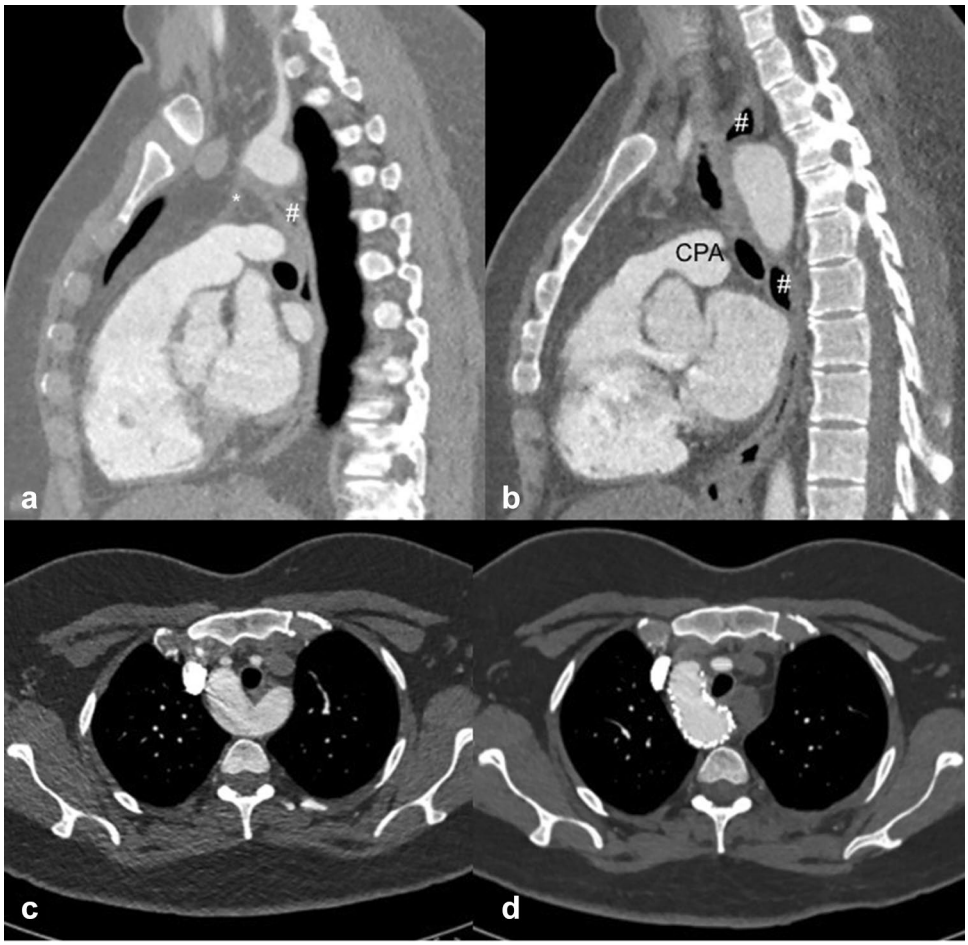

Fig. 1 Pre-operative chest computed tomography angiogram: **a** Sagittal reconstruction. **b** Sagittal reconstruction: compression of the oesophagus. **c** Axial cross-section: the oesophagus is compressed within the angle between the aortic arch and the Kommerell diverticulum. **d** Post-operative chest computed tomography angiogram after endovascular exclusion of the Kommerell diverticulum (*presumed location of the left ligamentum arteriosum; #Esophagus; *CPA* crossed pulmonary arteries)

ligamentum arteriosum, which connected the Kommerell diverticulum to the left pulmonary artery.

A three-stage surgical management was decided to exclude the aneurysm on one hand and to treat the oesophageal compression on the other hand.

The first procedure was a transposition of the LSA into the LCCA through a left transverse cervical approach. The second procedure consisted of the exclusion the Kommerell diverticulum by thoracic endovascular aortic repair (TEVAR). The third procedure was an aneurysmorrhaphy of the excluded Kommerell diverticulum and release of the oesophagus by division of the left ligamentum arteriosum through a left posterolateral thoracotomy. Because association with CPA reduced the available space to access the oesophagus thought of a left thoracotomy, dissection of the oesophagus was guided using a gastric catheter and carefully handled to prevent any vagus nerve injury, especially since unusual courses of recurrent laryngeal nerves have been described in case of right aortic arch [10]. The early postoperative course was uneventful. Dysphagia symptoms completely disappeared after surgery. Postoperative CTA showed a complete exclusion of the Kommerell diverticulum with no endoleak and confirmed the supra-aortic trunks' patency (Fig. 1d). After a 6-month follow-up, the patient was free from dysphagia and had regained 5 kg.

Discussion

Crossed pulmonary arteries are a rare and asymptomatic vascular anatomical variation. Consequently, in most of reported cases, it is usually diagnosed incidentally in association with a cardiac malformation. Its embryologic origin has not been clearly understood yet [4]. According to some authors, CPA is the result of a differential growth of the pulmonary trunk with an anticlockwise rotation of the branches of the pulmonary artery [9, 11]. It has been reported to be associated with various cardiac congenital malformations such as persistent truncus arteriosus, interrupted aortic arch, tetralogy of Fallot, atrial septal defect, left superior vena cava, and patent ductus arteriosus [1, 2, 8, 9]. CPA can also be associated with chromosomal abnormalities: trisomy 18, 22q11 deletions [9]. In our patient, CPA was incidentally discovered on preoperative CTA 3D reconstruction and the echocardiogram excluded any other cardiac congenital malformation, especially a patent ductus arteriosus.

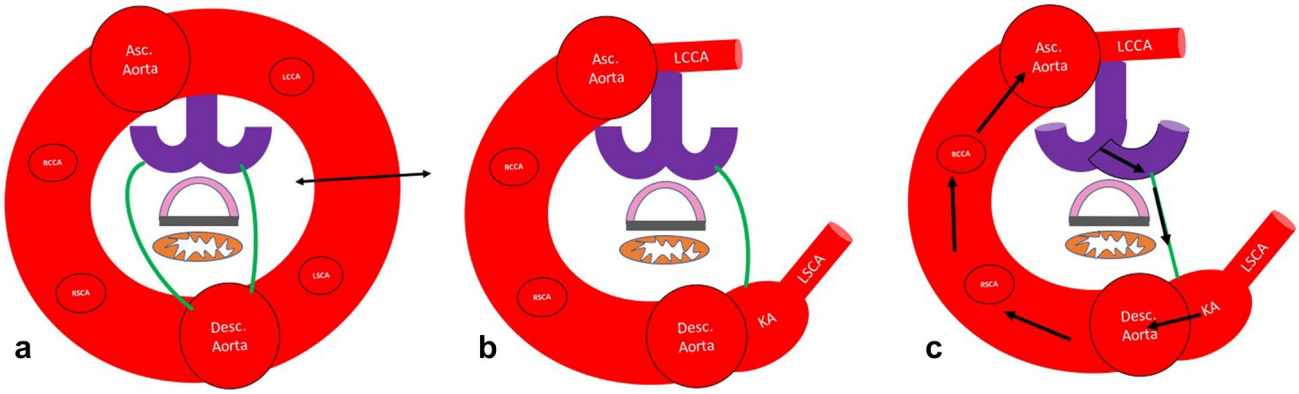

Fig. 2 **a, b** Formation of a right-sided aortic arch with Kommerell diverticulum and aberrant left subclavian artery associated with a left ligamentum arteriosum according to Corone and Vernant's theory with Neuhauser's anomaly; by interruption between left common carotid artery and left subclavian artery. **c** Vascular spiral pathway between crossed pulmonary arteries to the aortic arch (black arrows) (*Asc. Aorta* ascending aorta, *Desc. Aorta* descending aorta, *RCCA* right common carotid artery, *RSCA* right subclavian artery, *KA* Kommerell diverticulum)

Kommerell diverticulum is an aortic dilatation at the level of the origin of an aberrant subclavian artery. Although uncommon, this well-known anatomical configuration can lead to an oesophageal compression resulting in a disabling dysphagia. The compression is due to the retro-oesophageal position of both the diverticulum and the aberrant subclavian artery. Such an anatomical variation of the aortic arch can be explained by Edward's hypothesis of double arch [5] and Corone and Vernant's classification [3] (Fig. 2a).

Regarding the embryology, Kommerell diverticulum in a right aortic arch with an aberrant LSA is due to an interruption of the double aortic arch between the LSA and the LCCA. The LCCA arises from the proximal part of the right aortic arch as the first branch of the ascending aorta to be. The distal part of the left aortic arch persists posteriorly to the oesophagus as a diverticulum. In our case, this conformation was associated with a left-sided ligamentum arteriosum, as described by Neuhauser [6] (Fig. 2a, b). The left ligamentum arteriosum connects the left pulmonary artery to the diverticulum. Vascular structures seem to be organized as a constraining spiral around the oesophagus.

The left ligamentum arteriosum is a tight-and-narrow structure, enclosing the oesophagus and the trachea into the "vascular spiral". In case of CPA, the ligamentum arteriosum is connected to the superior part of the left pulmonary artery, higher than usual, reducing, thus, the size of the "vascular spiral" in a cranial plane (Fig. 2c).

In such great vessels' embryological conformation, late onset of symptoms is uncommon. Although we did not have any previous imaging to assess the evolution in the diverticulum size, we believed that it was an indirect evidence of a significant and recent diverticulum growth.

Whereas open repair of Kommerell diverticulum in a right aortic arch requires a cardiopulmonary bypass, endovascular exclusion allows a minimally invasive approach to prevent from rupture or dissection. In most cases, an additional cervical debranching is necessary to bypass the proximal LSA to prevent any retrograde flow into the aneurysm and to revascularize the superior limb [7]. Nevertheless, in some cases, uncompression of the Kommerell diverticulum with a stand-alone TEVAR might not be sufficient enough to improve dysphagia, because the oesophagus would be still tightly constrained by both the left ligamentum arteriosum and the aneurysm. In our case, opening the "vascular spiral" by dividing its fibrous part, i.e., the left ligamentum arteriosum, in addition to the exclusion of the Kommerell diverticulum, appeared to be mandatory to achieve a complete treatment.

Conclusion

Although dysphagia is a well-known complication of Kommerell diverticulum with persistent left ligamentum arteriosum in a right aortic arch, the association with CPA can explain even more severe symptoms. A complete preoperative anatomical analysis is mandatory to plan the appropriate surgical strategy both to exclude the aneurysm and to treat dysphagia.

Author contributions Protocol/project development: SEB and PVData collection or management: SEB, IBA, PJ, and JMAData analysis: SEB, IBA, JMA, PJ, and PVManuscript writing/editing: SEB and IBA.

Compliance with ethical standards

Conflict of interest All authors declare that they have no conflict of interest.

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