Kommerell Diverticulum: A Rare Cause of Dysphagia in a Child

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Keypoints

What is known:

- Kommerell diverticulum is a rare congenital aortic arch anomaly.
- When associated with right aortic arch and aberrant left subclavian artery, it can cause a vascular ring.

What is added:

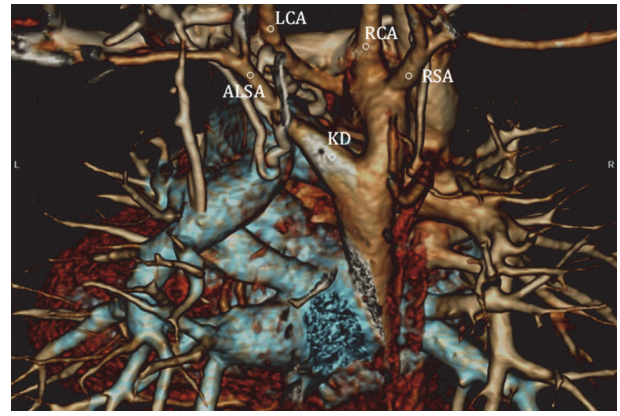
- Symptoms due to compression of the trachea and esophagus may present in pediatric age.
- A high suspicion is needed in children with symptoms that are difficult to interpret, but with known congenital heart disease associated with right aortic arch.

Introduction

An 8-year-old girl, with Down syndrome, a II/VI holosystolic murmur best heard at the apex, and mild dysphagia for solids was referred for cardiovascular consultation. The symptoms of mild dysphagia had started a year ago, intermittently, apparently without worsening through the year. The mother also reported rare episodes of wheezing, undocumented and without the need for specific treatment or hospitalization. The transthoracic echocardiogram revealed a small partial atrioventricular septal defect, a mitral valve cleft with moderate regurgitation, and a right aortic arch. A turbulent flow was also observed in the left subclavian region. Continuous Doppler interrogation revealed a gradient of 150 mmHg. An angio- thoracic tomography scan was carried out to clarify this finding. It revealed an aberrant left subclavian artery arising from a Kommerell diverticulum, and a small stenotic collateral blood vessel in the descending aorta close to the left subclavian artery (Fig. 1). The aberrant left subclavian artery had a retroesophageal course, compressing the upper third of the esophagus, which together with the Kommerell diverticulum, formed an incomplete vascular ring (Fig. 2). Kommerell diverticulum is a rare congenital aortic arch anomaly, with a reported incidence of 0.05%-0.1% in radiologic series.[1] It refers to dilation in descending aorta from which arises an aberrant subclavian artery, left or right, associated with both right and left aortic arch. The retroesophageal course of the aberrant subclavian artery is responsible for the compression of both the trachea and the esophagus, which potentially lead to dysphagia and respiratory distress.[2] Right aortic arch has an important association with congenital heart diseases, with a reported incidence of 75%-85% in types I and III right aortic arch and 5%-10% in type II.[3]

The surgical correction is recommended if the patient develop symptoms or if there is risk of rupture or dissection.[4-6] The majority of patients become symptomatic only in adult life.

After proper counselling, the parents decided for a wait-and-see approach, until further worsening of the dysphagia symptoms or need for concomitant mitral valvuloplasty when mitral regurgitation becomes severe



ALSA - aberrant left subclavian artery; KD - Kommerell diverticulum; LCA - left common carotid artery; RCA - right common carotid artery; RSA - right subclavian artery.

Figure 1. Angio-thoracic tomodensitometry with volumetric acquisitions. Tridimensional (3D) reconstruction. Posterior view.

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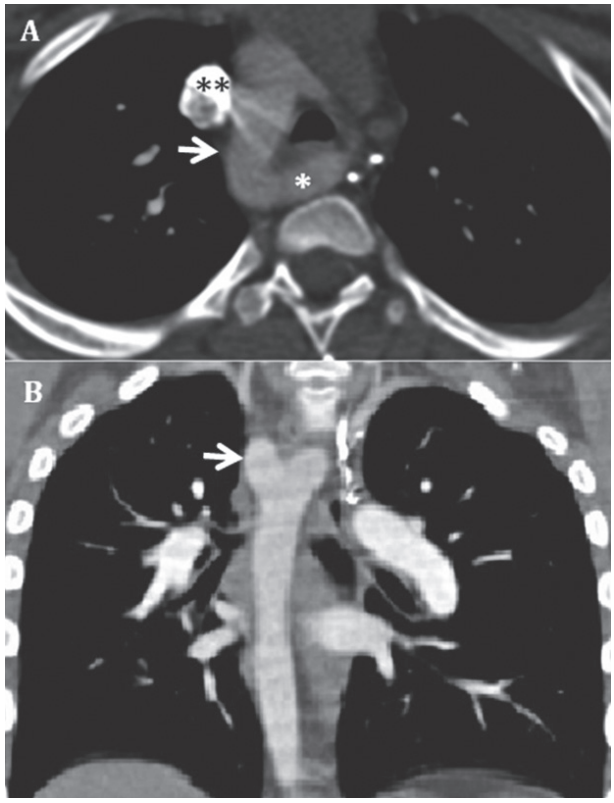

White arrow - Kommerell diverticulum; * - aberrant left subclavian artery; ** - superior vena cava.

Figure 2. Angio-thoracic tomodensitometry with volumetric acquisitions. A. Axial view. Note the incomplete vascular ring formed by the Kommerell diverticulum and the aberrant left subclavian artery, encompassing the trachea. B. Note the descending aorta right to the spine. Multiplanar reconstructions, coronal view.

Keywords: Aorta, Thoracic/abnormalities; Child; Deglutition Disorders/diagnosis; Diverticulum/congenital; Diverticulum/diagnostic imaging; Subclavian Artery/abnormalities; Subclavian Artery/diagnostic imaging; Vascular Ring/diagnostic imaging

Author Contribuitions

DF participated in the study conception or design. PVS, AF and AP participated in acquisition of data and in the analysis or interpretation of data. DF participated in the drafting of the manuscript. PVS, AF and AP participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki 2013).

Provenance and peer review

Not commissioned; externally peer reviewed

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data

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