Respir Case Rep 2022;11(2):93-97 DOI: 10.5505/respircase.2022.17362

Kommerell Diverticulum: A Rare Cause of Dyspnea and Dysphagia

Dispne ve Disfajinin Nadir Bir Nedeni Olarak Kommerell Divertikülü

Refika Hural[1], Gülsüm Bingol[3], Enis Öztürk[2], Ebru Serin[4], Seda Tural Onur[5], Emir Özgür Barış Ökçün[3]

Abstract

Kommerell's diverticulum (KD) is a congenital dilatation of the distal portion of the aortic arch that is usually located at the origin of an aberrant right subclavian artery (ARSA) or the aberrant left subclavian artery (ALSA). We describe here a 40-year-old male patient with a right-sided aortic arch (RAA) with ligamentum arteriosum (LA) and ALSA originating from the KD. The applied to our outpatient clinic with a history of dyspnea and dysphagia that had persisted for the last year. The patient underwent chest computed tomography (CT) in our clinic, revealing ALSA-level KD in RAA with ligamentum arteriosum. Surgical resection of KD and LA was planned based on the symptoms. NE was resected from the descending aorta and ALSA was transferred to the left carotid artery, and the patient was discharged 6 days after the operation without complications. After the operation, the patient's dyspnea and dysphagia subsided completely.

Key words: *Kommerell Diverticulum, Congenital Anomalies, ARSA, ALSA.*

Öz

Kommerell divertikülü (KD), genellikle aberran bir sağ subklavyen arterin (ARSA) veya aberran sol subklavyen arterin (ALSA) çıkışında bulunan aortik arkın distal kısmının konjenital dilatasyonudur. Bu olguda, sağ taraflı aortik ark (RAA) ile beraber ligamentum arteriyozumu (LA) olan ve KD'den kaynaklanan ALSA'sı olan 40 yaşında bir erkek hastayı tanımladık. Polikliniğimize başvurmuş bu hastanın son bir yıl içinde devam eden nefes darlığı ve yutma güçlüğü öyküsü vardı. Kliniğimizde hastaya, ligamentum arteriyozumlu RAA'da ALSA düzeyinde KD'ü göstermek için akciğer tomografisi çekildi. Semptomlar nedeniyle KD ve LA'nın cerrahi rezeksiyonu planlandı. İnen aortadan KD rezeke edildi ve ALSA sol karotid artere transfer edildi. Hasta ameliyattan 6 gün sonra komplikasyonsuz olarak taburcu edildi. Ameliyattan sonra hastanın dispne ve disfajisi tamamen kayboldu.

Anahtar Sözcükler: *Kommerell Divertikülü, Konjenital Anomaliler, ARSA, ALSA.*

[1]Department of Cardiology, Dr. Burhan Nalbantoğlu State Hospital, Nicosia, Cyprus
[2]Bakırköy Prof. Mazhar Osman Research and Training Hospital for Neurologic and Psychiatric Diseases, İstanbul, Türkiye
[3]Department of Cardiology, Memorial Bahçelievler Hospital, İstanbul, Türkiye
[4]Department of Cardiology, İstanbul University Cerrahpaşa, Institude of Cardiology, İstanbul, Türkiye
[5]Department of Chest Diseases, Yedikule Chest Diseases and T horacic Surgery Training and Research Hospital, İstanbul, Türkiye

[1]Dr. Burhan Nalbantoğlu Devlet Hastanesi, Kardiyoloji Bölümü, Lefkoşa, Kıbrıs
[2]Bakırköy Prof. Dr. Mazhar Osman Ruh Sağlığı ve Sinir Hastalıkları Eğitim ve Araştırma Hastanesi, Radyoloji Anabilim Dalı, İstanbul
[3]Memorial Bahçelievler Hastanesi, Kardioloji Bölümü, İstanbul
[4]İstanbul Üniversitesi Cerrahpaşa, Kardiyoloji Enstitüsü, Kardioloji Bölümü, İstanbul
[5]Yedikule Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim Araştırma Hastanesi, Göğüs Hastalıkları Bölümü, İstanbul

Submitted (Başvuru tarihi): 08.02.2022 ***Accepted (Kabul tarihi):*** 30.03.2022

Correspondence (İletişim): Gülsüm Bingol, Department of Cardiology, Memorial Bahçelievler Hospital, İstanbul, Türkiye

e-mail: bulut_gulsum@hotmail.com

Kommerell diverticulum (KD) is congenital dilatation of the distal part of the aortic arch that is usually found at the origin of an aberrant right subclavian artery (ARSA) or aberrant left subclavian artery (ALSA) (1). It was first described in 1937 by B. F. Kommerell, whose definition related to a patient with ARSA and left aortic arch (LAA) (2). Although most patients with KD are asymptomatic, dilatation of the diverticulum may compress the trachea or esophagus, causing such symptoms as dysphagia, dyspnea, wheezing, coughing and chest pain. The presence of an abnormal subclavian artery or vascular ring may worsen compression symptoms (2,3). Esophageal and tracheal occlusions are well-known indications for the surgical treatment of KD (4).

CASE

We describe here the case of a 40-year-old man who was admitted to our outpatient clinic with a right-sided aortic arch (RAA) with a ligamentum arteriosum (LA) and an ALSA arising from a KD. The patient had a history of dyspnea and dysphagia lasting for the last one year, as well as hypertension and diabetes mellitus. A physical examination revealed blood pressure of 120/70 mmHg that was the same in both arms, and weak left brachial and radial artery pulses.

A chest X-ray revealed an enlarged artery in the proximal descending aorta. Laboratory results were within normal limits. Transthoracic echocardiography showed normal left ventricular systolic function. CT was performed to demonstrate KD at the level of the ALSA in the RAA with a ligamentum arteriosum (Figure 1). The origin of ALSA, which causes significant compression by the LA (not demonstrated in the figures) and RAA in both the esophagus and trachea, has been shown to be aneurysmal (Figure 1 and 2).

Surgical resection of the KD and LA was planned based on the symptoms. The LA between the origin of the descending aorta and the left pulmonary artery was separated. The KD was resected from the descending aorta and the ALSA was transferred to the left carotid artery. The patient was discharged 6 days after the operation without complication. The dyspnea and dysphagia subsided completely after the operation. Postoperative CT images are presented in Figures 3, 4 and 5.

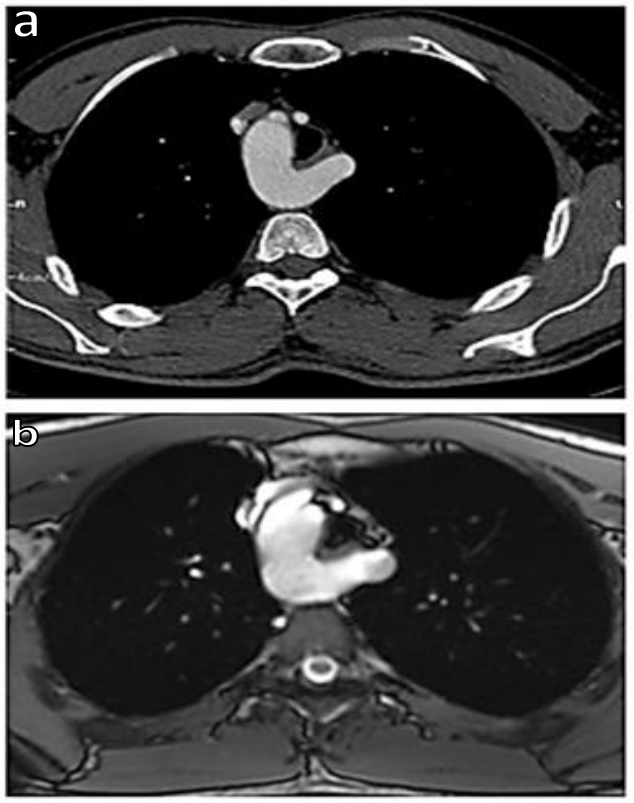

Figure 1: Axial CT (a) and corresponding magnetic resonance (MR) images (b) show the RAA and KD causing mild compression of the trachea and prominent compression of the esophagus

Figure 2: Coronal CT showing the ALSA arising from KD

DISCUSSION

The prevalence of congenital aortic arch anomalies ranges from 1–2% in the general population (5). These anomalies are commonly asymptomatic, and can be left/right-sided, double (DAA) or cervical aortic arch (CAA). Regardless of which, abnormal isolations of the

subclavian, brachiocephalic or carotid arteries and vascular rings may produce symptoms (6).

A normal LAA is generated by the regression of the right canal, RAA and the right dorsal aorta. The first branch of a normal LAA is the right brachiocephalic artery, followed by the left common carotid and left subclavian arteries. Furthermore, the first branch of a normal LAA is the right brachiocephalic artery, followed by the left common carotid and left subclavian arteries. The prevalence of RAAs ranges from 0.05–0.1% of the population (7), while DAAs and CAAs are rarer vascular anomalies.

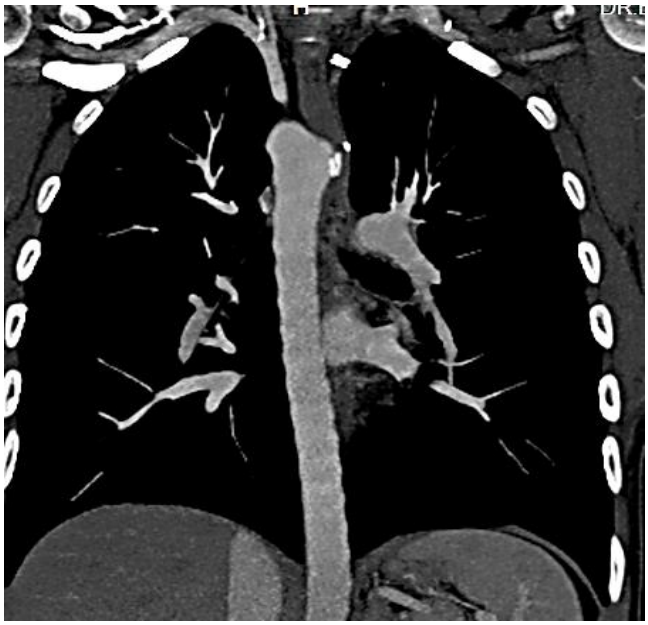

Figure 3: Post-operative coronal CT image showing metallic clips at the site of the resected KD

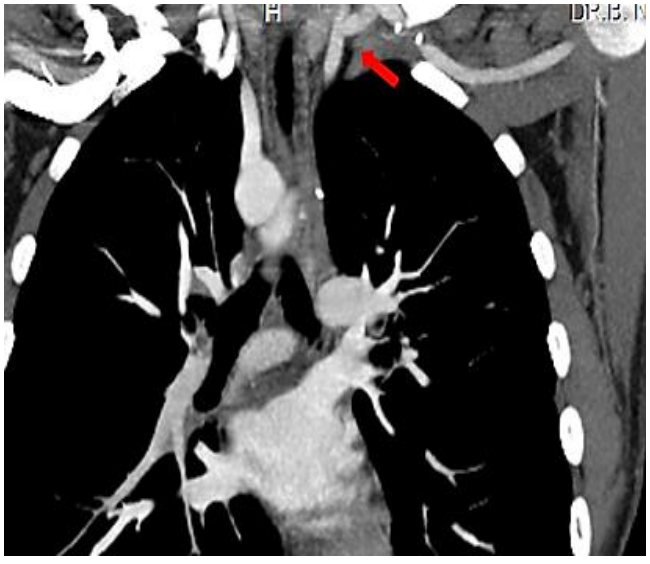

Figure 4: Postoperative coronal CT image showing a carotico-subclavian by-pass

According to the branching pattern of great vessels referred to by Bravo et al. (5), the LAA can divide into three groups as ARSA, right subclavian artery isolation and normal. Furthermore, RAAs can be divided into three types based on the branching pattern of the arch veins as left subclavian artery isolation, mirror image branching and ALSA (6).

In the study by Türkvatan et al. (8), one of the most common congenital anomalies of the aortic arch was stated to be aberrant subclavian artery, whereas the incidence of ARSA resulting from normal LAA is in the 0.4–2.3% range in the general population. Yang et al. (9) reported that ALSA originating from the RAA is rare, with an incidence of only 0.05% in the general population.

KD is an aneurysmal dilatation of the distal part of the aortic arch or at the origin of an aberrant subclavian artery (4). KD is widely thought to result from degeneration associated with atherosclerosis, or to be congenital (10). It has also been reported that this condition, which is more common after the age of 50, has no gender predominance. According to Kommorell's initial definition (2), the KD consists of an abnormal artery originating from the left aortic arch, while the right subclavian artery emerges as the last branch of the aortic arch. This branch then passes from the proximal descending aorta behind the esophagus to the right arm.

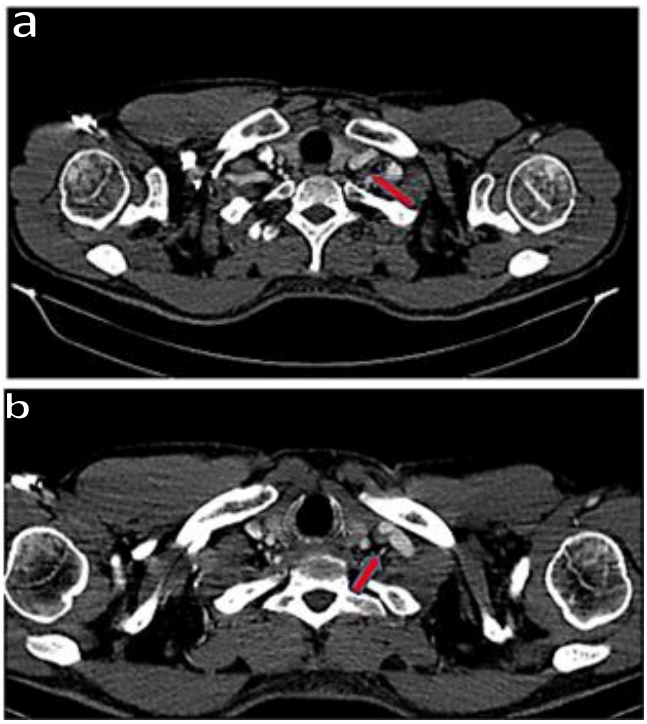

Figure 5: Postoperative axial CT images showing a carotico-subclavian by-pass

Although the existence of seven types of KD has been reported in literature, three are more common: (a) LAA diverticula with ARSA, (b) RAA diverticula with ALSA, and (c) diverticula at the aortic-ductal junction (4,8). Patients are usually asymptomatic, but may experience trache-oesophageal compression, although dilation of the KD can sometimes lead to such symptoms as shortness of breath, wheezing, coughing or chest pain due to the compression of the trachea or esophagus (2,4,11).

Vascular rings are rare congenital anomalies of the aortic arch and its branches in which the trachea and esophagus may be compressed by a combination of LA and an abnormal aortic arch course. The RAA is the second most common form of a complete vascular ring formed by a patent duct or ligamentum arteriosum, contralateral to the ascending aorta (12). Different imaging methods, such as echocardiography, barium swallow, bronchoscopy, esophagography, angiography, CT and magnetic resonance imaging (MRI) are available for the diagnosis of this anomaly (11). CT and MRI are considered the best diagnostic approaches to KD. The diagnosis of our case was confirmed by CT angiography, which shows the RAA with the ALSA and KD.

Complications of KD can be serious, with diverticulum rupture or dissection reported in patients with aberrant subclavian arteries (13). Symptomatic patients should be considered candidates for surgery when the minimum size of the KD is greater than 2 cm or the aneurysm diameter exceeds 1.5 times that of the associated subclavian artery surgery (14).

Although surgical interventions in asymptomatic patients is controversial, some authors recommend surgery in such patients due to the risk of rupture or dissection (3). Various surgical techniques are available in this regard, such as simple ligation of the left subclavian artery, resection of the KD, transplantation of the left subclavian artery to the left carotid artery, and endovascular repair (4,11).

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Planning and Design - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Supervision - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Funding -; Materials - R.H., E.Ö.; Data Collection and/or Processing - R.H., E.Ö.; Analysis and/or Interpretation - G.B., S.T.O.; Literature Review – E.S., G.B.; Writing - G.B., R.H., E.Ö.B.Ö.; Critical Review - S.T.O., E.Ö.B.Ö., G.B.

YAZAR KATKILARI

Fikir - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Tasarım ve Dizayn - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Denetleme - R.H., G.B., E.Ö., E.S., S.T.O., E.Ö.B.Ö.; Kaynaklar -; Malzemeler - R.H., E.Ö.; Veri Toplama ve/veya İşleme - R.H., E.Ö.; Analiz ve/veya Yorum - G.B., S.T.O.; Literatür Taraması - E.S., G.B.; Yazıyı Yazan - G.B., R.H., E.Ö.B.Ö.; Eleştirel İnceleme - S.T.O., E.Ö.B.Ö., G.B.

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