Syncope as Initial Presentation of Kommerell Diverticulum

Ronny Cohen, M.D., F.A.C.C.^{1,2} Pablo Loarte, M.D.² Christine Garcia, M.D., M.P.H.³
Lizmer Diaz, M.D.² Brooks Mirrer, M.D.^{1,2}

¹ Division of Cardiology, Department of Medicine, NYU School of Medicine, Brooklyn, New York

² Division of Cardiology, Department of Medicine, Woodhull Medical Center, Brooklyn, New York

³ Division of Cardiology, Department of Medicine, St. George's University School of Medicine, St. George's University, University Centre, Grenada, West Indies.

Address for correspondence and reprint requests Ronny Cohen, M.D., F.A.C.C., Woodhull Medical Center, NYU School of Medicine, 760 Broadway, Brooklyn, NY 11206 (e-mail: ronny.cohen@nychhc.org).

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Abstract

Keywords

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Kommerell diverticulum (KD) is an aortic arch diverticulum at the origin of an aberrant subclavian artery. It is a rare anatomical anomaly that can be associated with a double aortic arch, a left aortic arch, and anomalous origin of the right subclavian artery or a right aortic arch with anomalous left subclavian artery. We present a case of KD presenting initially as syncope, as well as a review of the literature of this rare syndrome, including diagnosis, imaging techniques, and current surgical treatments.

Kommerell diverticulum (KD) is an aortic arch diverticulum at the origin of an aberrant subclavian artery. Dr. Friedrich Kommerell first described this syndrome in 1936, as a diverticular outpouching at the origin of an aberrant right subclavian artery with a left-sided aortic arch.^{1,2} KD is a rare anatomical anomaly that can be associated with a double aortic arch, a left aortic arch with anomalous origin of the right subclavian artery (ARSA) or a right aortic arch (RAA) with anomalous left subclavian artery (ALSA).³ There are few reported cases in the literature, and the clinical presentation is variable and nonspecific.

Case Report

A 47-year-old woman with history of hypertension presented to the emergency department complaining of an acute syncopal episode. She was in bed and stood up, then suddenly collapsed after a few steps. She had a momentary loss of consciousness. She recalled that initially, she felt dizzy, described as “light-headedness for a few seconds like everything around her was moving with a sensation of weightless in her body.” She then passed out shortly after. As per her relatives who witnessed the incident, the episode lasted for less than 1 minute. There was no past history of syncope. Her past medical history is significant for hypertension for 8 years and dyslipidemia. Her current medications are Altace (Ramipril) 5 mg once daily, Metoprolol

succinate ER (Metoprolol) 50 mg twice daily, and Zocor (Simvastatin) 40 mg once daily. Her vitals were stable at presentation. She was afebrile, her heart rate was 86 bpm, respiratory rate was 14 breaths per minute, and blood pressure was 145/85 mm Hg. There was no presence of orthostasis. Pulses were 2+, equal in all four extremities.

On physical examination, there were no neurological findings and cardiovascular system was normal. Cardiac auscultation examination with normal S1 and S2, no S3 or S4 with a regular rate and rhythm. There was no PMI displacement and no carotid bruits. No murmurs, rubs, or gallops were noted on exam. Lung examination revealed clear auscultations and percussion in all lung fields, no wheezing, rales, or rhonchi. Abdomen was soft, nontender with normal bowel sounds, no bruits on auscultation, no hepatomegaly, and spleen was not palpable. Extremities were without deformities or joint abnormalities, no edema present, and peripheral pulses were intact. Neurological exam showed intact cranial nerves II to XII, strength and sensation symmetrically throughout, reflexes were 2+ throughout, and cerebellar testing was unremarkable.

Laboratory evaluation did not show any abnormalities; cardiac biomarkers were negative and hemoglobin was within normal limits. The electrocardiogram showed sinus rhythm with borderline left axis deviation, poor R wave progression in anterior leads, and diffuse nonspecific T wave abnormalities (►Fig. 1).

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Figure 1 Patient's electrocardiogram on arrival to emergency room.

Head computed tomography (CT) did not show any mass, hemorrhage, or infarction and chest radiograph incidentally showed a RAA (►Fig. 2).

Echocardiogram revealed normal left ventricular systolic function with an ejection fraction of 63%, dilated left atrium, and right ventricle. Chest CT was ordered for the abnormal radiographic finding. The study identified a right-sided aortic

arch with a dilated retroesophageal vascular diverticulum causing a mass effect in the esophagus and a left subclavian artery arising from the diverticulum (►Figs. 3 to 5). The size of the diverticulum was 47 mm in its larger diameter and the

Figure 2 Chest radiograph with a right-sided aortic arch.

Figure 3 Anteroposterior angiogram, showing a left subclavian artery (arrow A) arising from a hidden Kommerell diverticulum, ascending aorta (arrow B) and superior vena cava (arrow C).

Figure 4 A posterior view of descending aorta, we can see the aberrant left subclavian artery (arrow A and B) arising from the Kommerell diverticulum (arrow C) and traveling to the left side. Descending aorta (arrow D).

Figure 5 Axial view of chest computed tomography showing a Kommerell diverticulum (arrows A and B) surrounding partially the esophagus from behind (arrow C).

descending aorta was following a course from the right to left side when approaching the abdominal cavity.

Surgical repair of the anomaly was an option offered to the patient. However, after full discussion of the benefits and risks, she opted for conservative management despite the risk of complications.

Discussion

Embryologically, the KD represents the persistent distal end of an interrupted fourth aortic arch between the carotid and subclavian arteries. An additional definition of KD is the one that describes it as a saccular aneurysmal dilation at the origin of an ARSA or ALSA.³ Felson and Palayew anatomically classified KD with right-sided aortic arch aneurysm and divided in two types; being the type 2, the most common.^{4,5} Another proposed classification is Edward's classification and describes 3 types of RAA.⁶ The presence of a left aortic arch and ARSA has been found in 0.5% of the population. In our case, we have a KD with ALSA, a very rare finding. An ALSA is found only in ~0.1% of the population and is the most commonly anomaly in patients with RAA.^{3,7} Only 60% patients with an aberrant subclavian artery have a KD.⁷ The presence of an RAA has been documented to occur in only 0.05% of the population.⁸ Aberrant right subclavian arteries can traverse behind the esophagus upward to the contralateral side, going either through the retroesophageal space (80%), between esophagus and trachea (15%) or anterior to the trachea (5%).⁹ In our case, we have an aberrant LSA (Left Subclavian Artery) and as per literature, the course can vary; posterior or anterior to esophagus or anterior to both, trachea and esophagus.¹⁰

Diagnosis of KD

The diagnosis of KD can be made incidentally depending on the presenting symptoms. Most of the patients are asymptomatic and the presence of symptoms is related to the compression of surrounding structures. Patients can present with dysphagia, known as *dysphagia lusoria*, defined as "dysphagia by a trick of nature." When the esophagus is affected, dyspnea, wheezing, stridor, cough, recurrent pneumonia or chest pain occur.¹¹ There are reported cases in which patients present with aortic aneurysm-like symptoms and a KD dissection was an incidental finding.^{12–14} Syncope is not a symptom of presentation in a patient with KD and has not been previously reported. There was one case in the literature where the patient had a similar anatomic anomaly with a history of three previous syncopal episodes.¹⁵ The syncopal episode in our case may likely be attributed to a transient subclavian steal-like syndrome caused by compression of the aberrant subclavian by any adjacent structure. Unfortunately, there is no previously explained mechanism for the syncope, the only suggestion is the possibility of a subclavian steal syndrome or phenomenon caused by compression of the aberrant subclavian artery, stenosis, or intra-arterial high pressures. Unfortunately, the CT angiogram was not able to evaluate the complete course of the subclavian to provide anatomical explanation for this finding. Syncope may be an incidental finding. Orthostatic hypotension excluded due to patient's age, lack of reproducibility in the emergency room, and elevated blood pressure on presentation.

Several imaging techniques can be used to confirm the diagnosis of KD. Indirect evidence of possible vascular anomaly can be made by showing indentation of the esophagus or compression of the upper airways. This can be demonstrated

through barium swallow, esophagogastrosocopy, and bronchoscopy. Actually the most widely used and accurate imaging studies are the computerized axial tomography angiography and the magnetic resonance angiography.^{4,16}

The major complications when a KD exists are related to the presence of aneurysm of aorta or when the KD diameter is large. Aneurysm has been observed in 3 to 8% of the patients with aberrant subclavian artery. The risk for rupture or dissection is variable in the literature, ranging from 19 to 53% in some of the case report series.¹⁶ Presence of an aneurysm has been associated with rupture or dissection, which can result in patient death. The mean size associated with rupture was 5.8 cm in one case series report.^{16,17} Another serious complication is embolization.⁸

Surgical Management of KD

Standard surgical management has not been established yet due to the rarity of this condition. In one study of 10 cases of KD with concurrent aneurysmal degeneration of the adjacent descending aorta, a graft was used to repair the subclavian artery or correct the thoracic aneurysm. From this group of patients, only two had a left-sided aortic arch and underwent graft plus repair of the ALSA. Four patients died in the postsurgical follow-up, however, these patients were not the best surgical candidates due to age and/or concurrent malignancy. One patient had a chylothorax and died of multiorgan failure after undergoing surgery for repair.¹⁸ The mortality from a surgical approach can be as high as 16.6%.⁷ The surgical approach varies according to the anatomy of the anomaly and this can influence also in the mortality rate.

Due to the rareness of this condition and other superimposed-associated anomalies, there is no standard indication for surgical management, either in symptomatic or asymptomatic patients. In one review of 32 previously published cases of RAA with aneurysm, aggressive surgical management was recommended for aneurysms above 3 cm, due to the complexity of large KD with aneurysm.¹⁶ In a recent study, surgical repair was recommended in symptomatic aneurysm of 5 cm or greater.^{7,8} Our patient would likely have benefited from an elective surgical repair since the diameter was close to 5 cm. We recommend assessing every patient with KD putting special attention to the size of the lesion and persistence of symptoms. The chances of rupture were more common in lesions equal or above 50 mm with or without symptoms.

In one study, six patients underwent an open surgical repair (thoracotomy, either left or right). All procedures were two-stage repairs, using a graft to repair the aorta or the subclavian artery, approaching the aorta in the first stage, then approaching the aberrant subclavian artery.^{7,8} This approach had zero mortality with only one case developing mediastinitis immediately postoperation. There were no complications in the follow-up period. Other described surgical approach is the hybrid, which combines an endovascular repair plus an open surgical repair.¹⁴ In one case report, the transverse diameter of the KD was 3.3 cm. The patient underwent thoracic aortic endovascular repair (TEVAR) plus use of graft to repair the aberrant subclavian artery.¹⁷

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