GACETA MÉDICA DE MÉXICO

CLINICAL CASE

Kommerell's Diverticulum (KD)

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Abstract

Right aortic arch with aberrant left subclavian artery from a Kommerell's diverticulum is a very rare variant of the incomplete vascular ring. Associated symptoms are caused due to tracheal or esophagus compression. Magnetic resonance is the gold standard for diagnosis. Surgical treatment is recommended for symptomatic patients or asymptomatic patients with a large diverticulum. We report three consecutive cases of patients with Kommerell's diverticulum, aberrant left subclavian artery, and right-sided aortic arch. (Gac Med Mex. 2016;152:382-6)

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KEY WORDS: *Kommerell's diverticulum. Aberrant left subclavian artery. Vascular ring.*

Introduction

The term vascular ring refers to an anomaly of the aortic arches where the trachea and the esophagus are surrounded by these structures. It accounts for less than 1% of all congenital heart anomalies. Its classification depends on the arch's position with regard to the trachea (Table 1). Within the trachea, the right aortic arch with aberrant left subclavian artery (ALSA) is found with left ligamentum arteriosum, which shows an incidence of 0.05%. In this right aortic arch anomaly, the left carotid is originated in the first place, followed by the right brachiocephalic trunk and, finally, the left subclavian artery, which arises from a diverticulum of the descending aorta, known as Kommerell's diverticulum (KD), with retro-esophageal trajectory, and with ligamentum arteriosum taking place at its base[1-3].

The purpose of this article is to present three clinical cases of a vascular ring rare variant with right aortic arch.

Case 1

Two-year old male patient with a history of laryngeal stridor since birth, which was exacerbated with feeds, chronic cough, two episodes of bronchiolitis and one of community-acquired pneumonia. Performing a bronchoscopy was decided, with a pulsatile mass being found, which was suspected to be a vascular ring. At the moment of examination in our department, the patient had no cardiovascular symptoms, with Ross functional class I, and no relevant findings on physical examination. Chest X-ray showed right superior mediastinum broadening with image suggestive of right aortic arch. The echocardiogram showed no intracardiac defects. A heart and larger arteries angiotomography

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Table 1. Congenital anomalies of the aortic arch classification and supra-aortic trunks anatomical variations

Congenital anomalies of the aortic arch
– Left aortic arch with aberrant right subclavian artery – Right aortic arch with mirror image branching – Right aortic arch with aberrant left subclavian artery – Cervical aortic arch – Interrupted aortic arch
Supra-aortic trunks anatomical variations
– Brachiocephalic arterial trunk and common right left artery common origin Common left carotid artery origin in the brachiocephalic arterial trunk – Aberrant left vertebral artery (aortic arch-origin)

was performed, where a right aortic arch surrounding the right lateral edge of the trachea was found, with no evidence of compression of the latter. Images of a right aortic arch and a KD, out of which the left subclavian was arising, were observed in the tridimensional reconstruction (Fig. 1).

The patient continued with respiratory symptoms, inspiratory stridor and tracheal cough. The case was consulted with the cardiovascular surgery department, arriving to the conclusion that he was candidate for surgical treatment. Section and suture of the vascular ring was practiced, in addition to aortic arch-pexy without complications; the patient is currently asymptomatic.

Figure 1. *Computed angiotomography 3D reconstruction. The view is oblique, and the posterior side of the heart is identified, as well as the trajectory to the right of the thoracic aorta. The origin of the subclavian artery is appreciated in the posterior position and as the last vessel of the aortic arch, which originates from a Kommerell's diverticulum.*
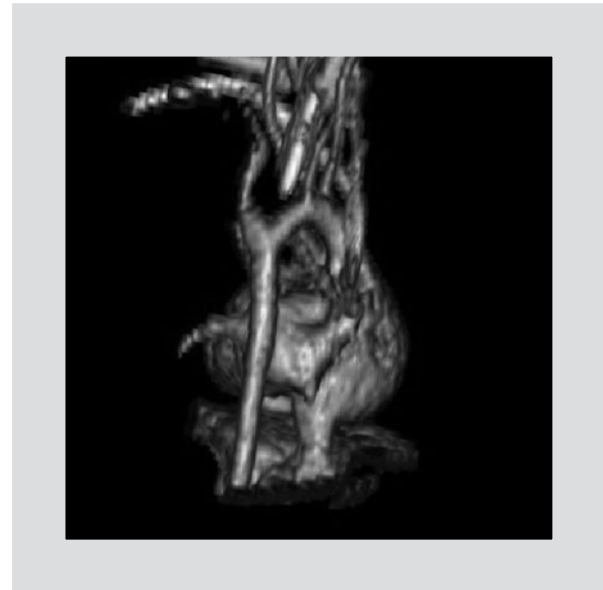

Figure 2. *Computed angiotomography 3D reconstruction. The view is posterior, and the posterior side of the heart is identified, as well as the trajectory to the right of the thoracic aorta. The origin of the subclavian artery is appreciated in the posterior position and as the last vessel of the aortic arch, which originates from a Kommerell's diverticulum.*

Case 2

Preterm male patient of 28 weeks' gestation who was referred to the neonatal intensive care unit where, at 23 days of life, he displayed clinical data consistent with patent ductus arteriosus with hemodynamical repercussion. On physical examination, a grade 2/6 continuous murmur with predominance at the second left intercostals space was found, as well as widened pulses and wide differential blood pressure. The echocardiogram revealed the presence of a right aortic arch and patent ductus arteriosus. Surgical closure of the ductus was performed, with right aortic arch, 10-mm long ductus arteriosus and left subclavian anomalous origin intraoperatively found. The ductus was ligated after the left subclavian artery emergence, with desaturation and arrhythmia occurring at that moment, which prompted giving up the ductus closure. In view of the surgical findings, a heart and larger arteries angiotomography was performed, with a KD, ALSA and left aortic arch being found (Fig. 2). Maintaining the patient under surveillance was decided, and only in case of vascular ring-associated symptoms would surgical management be considered again. The patient contracted nosocomial sepsis as a complication and died.

Case 3

Male patient of 3 years and 11 months of age with a history of vaginal birth, 1,900 g weight, 46 cm length, cried and breathed at birth, with sepsis ongoing at that moment. A cardiac murmur was detected at 15 days of life, with congenital heart disease without hemodynamical repercussion being suspected. He was referred to our department at 11 months of age, where he was found free of cardiovascular symptoms, with Ross functional class I and with low left parasternal holosystolic murmur, grade 3/6, with band-like radiation. The echocardiogram revealed a 5-7-mm perimembranous interventricular communication, left-to-right shortcut with a 60 mmHg gradient throughout, pulmonary artery systolic pressure (PASP) of 30 mmHg, estimated by gradient though the interventricular defect, pulmonary artery left branch with an unusual origin and 2 x 3 mm ductus arteriosus with left-to-right shortcut. In view of the echocardiographic image of the pulmonary branches, a heart and larger arteries angitomography was performed, where right aortic arch, KD, ALSA and 4.4-mm interventricular communication were reported (Fig. 3). The patient was seen again as an outpatient 3 years later (he was lost to follow-up due to extra-medical reasons) and was found at Ross functional class I, with grade 3/6 low left parasternal holosystolic murmur, chest X-ray with slight cardiomegaly, increased pulmonary flow with right predominance; echocardiogram demonstrated 6.4 mm perimembranous interventricular communication and another 3-mm mesotrabecular, left-to-right shortcut with 37 mmHg gradient throughout, PASP of 63 mmHg, right aortic arch, 4 mm ductus arteiosus with 9 mmHg gradient throughout. The case was presented in medical-surgical session, and interventricular communication closure and ductus arteriosus ligation was decided. Closure of the ventricular septal defect was performed with a Teflon patch and double ligation, as well as closure of the foramen ovale without intraoperative or postoperative complications. The control echocardiogram showed no residual defects, with 20 mmHg PSAP inferred by the tricuspid insufficiency. At follow-up as an outpatient, the patient remains asymptomatic, with Ross functional class I and with no respiratory symptoms present.

Discussion

In 1735, David Bayford first described the right aortic arch with aberrant right subclavian artery anomaly.
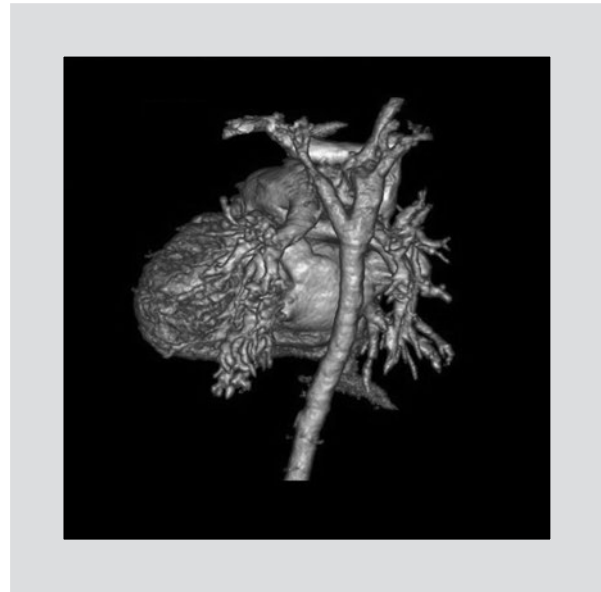

Figure 3. *Computed angiotomography 3D reconstruction. The view is oblique, and the posterior side of the heart is identified, as well as the trajectory to the right of the thoracic aorta. The origin of the subclavian artery is appreciated in the posterior position and as the last vessel of the aortic arch, which originates from a Kommerell's diverticulum.*

In 1936, Burckhard Kommerell determined for the first time the clinical presentation in a patient who underwent an esophagogram that showed posterior compression of the esophagus upper third originated by a pulsatile mass, describing in the patient's autopsy what now is known as KD. In 1988, Felson divided the entity in 3 categories: normal KD, aneurismatic dilatation of the diverticulum and aneurism of the distal aberrant subclavian artery[4].

During aortic arch embryologic development, there are 6 pairs of aortic arches. In normal growth, the distal part of the first and second aortic arches disappears, and the proximal part forms the hyoid and maxillary arteries. The third pair of arches forms the internal carotid arteries' proximal part. The fourth left ventral aortic arch will form the aortic arch between the common carotid and the left subclavian artery, whereas the proximal portion of the fourth right aortic arch forms the proximal part of the right subclavian artery, the distal part of the right aortic arch between the seventh intersegmental artery undergoes regression, which determines the formation of the left aortic arch. The dorsal aorta distal to the sixth aortic arch forms the descending aorta distal to the left subclavian artery. The left subclavian artery and the distal portion of the right subclavian artery originate in the seventh arch of the dorsal aorta. The fifth aortic arch undergoes bilateral regression and

the sixth aortic arches originate the proximal portion of the pulmonary arteries and the left and right ductus arteriosus, with the right one normally disappearing and the left one remaining permeable during fetal life. If the fourth right aortic arch remains and the fourth left aortic arch between the common carotid and the left subclavian artery undergoes an anomalous regression, a right aortic arch will form, with an ALSA originating from a diverticulum in the junction of the aortic arch to the right and the right descending aorta, and obliquely passing in an upwards direction, behind the esophagus, towards the left arm. The diverticulum is generally well developed, since the fetal ductus arteriosus, in the ALSA origin, carries a large volume of blood[1,3,5].

Most patients are asymptomatic and there is no particular association with other cardiac anomalies. Some authors have reported ventricular septal defect as the most common associated malformation, while other series mention aortic coarctation, tetralogy of Fallot and larger arteries transposition. Symptoms related to the presence of a vascular ring may exist, which will depend on the degree of compression produced; symptoms will generally appear before the first month of age, with most part of them consisting in stridor, feeding difficulties, cough and sibilance. In adults, symptoms are driven by atherosclerotic changes, anomalous vessels rigor, dissection and aneurism with compression of adjacent structures causing dysphagia (dysphagia lusoria), dyspnea and chest pain[3,6,7].

Angiotomography is a non-invasive method that enables KD diagnosis; images can be reconstructed and seen from different angles, making it easier to locate and measure the size of the diverticulum. In addition, it shows the relationships of the diverticulum with adjacent structures and can be used to detect tracheal and esophageal compression[8].

According to angiotomographic findings, 4 different types of KD are described. The most common is a KD arising from a right aortic arch with ALSA; in these cases, the aberrant subclavian artery crosses behind the esophagus (80% of cases), but it can pass between the esophagus and the trachea (15% of cases) or anterior to the trachea (5% of cases). Occasionally, a left ligamentum arteriosus between the left subclavian artery and the left branch of the pulmonary artery forms a vascular ring. This ring is usually loose and doesn't cause tracheal or esophageal compression severe symptoms. The second type of KD occurs with a left aortic arch with an aberrant right subclavian artery; however the aberrant right subclavian artery not always arises from the aortic diverticulum. The ligamentum

arteriosus is located at the left side and does not produce a vascular ring. The third type is located at the aortic-ductal junction and appears as a protuberance in the internal part of the aortic isthmus, distal to the subclavian artery. Although 33% of infants show dilatation at the ligamentum arteriosus site, this dilatation resolves within the first months after birth. In some cases, the dilated diverticulum persists until adulthood, usually not accompanied by other aortic arch anomalies. The fourth type occurs with left aortic arch and right descending aorta with aberrant right subclavian artery[5,8,9].

Magnetic resonance imaging is the non-invasive study of choice since it provides optimal information, showing the anatomical characteristics of the ring and determining the surgical approach without the patient being exposed to radiation[3,4].

Surgical treatment is recommended in symptomatic cases, when signs and symptoms of airway or esophageal compression are present and in asymptomatic cases showing KD dilatation. Surgical approach can be with open surgery, endovascular therapy or using hybrid treatments. Their objective is KD resection and subclavian artery reconstruction in order to prevent ischemia of the arm and vertebrobasilar territory and subclavian artery steal syndrome[3,4,8]. In our case series, only one patient underwent KD resection owing to the persistence of respiratory symptoms secondary to extrinsic compression of the airway. In the second case, KD correction was deferred due to intraoperative hemodynamical instability, with the patient dying because of nosocomial sepsis. In the last of our cases, no KD resection was practiced owing to the absence of respiratory symptoms, which made for surgical procedure to be deemed unnecessary.

Endovascular aortic reparation associated with aortic branches re-anastomosis or subclavian artery bypass graft has been used in some cases, with decreased morbidity and mortality when compared with open surgery. The presence of ALSA requires proximal ligation and embolization with a coil device or some other occluding device to prevent type II internal leakage. Endovascular therapy with endovascular stent graft to exclude ALSA/KD, with distal ligation of the left subclavian artery and upper limb revascularization, is becoming the standard of care[10,11]

Conclusions

KD is a rare anomaly of the aortic arch that occasionally generates tracheal or esophageal compression symptoms. Knowledge of this pathology is important in order to enable opportune diagnosis and suggest a management plan.

Acknowledgements

Financial support

Conflicts of interests

There are no conflicts of interests related to the procedures involved with this publication.

Ethical standards

Informed consent was requested from the patients' parents for the publication of these cases.

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