

Atypical presentation of Kommerell's diverticulum

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The authors report a case of a 65-year-old male patient who, after complaint about retrosternal discomfort and going through a thoracic radiography with mediastinal enlargement, was submitted to magnetic resonance angiography, which detected a right aortic arch with aberrant left subclavian artery and Kommerell diverticulum. A literature review was the basis for our clinical conduct, for the surgical treatment is complex. The patient presented improvement with clinical treatment and, currently, is under ambulatory follow-up care, with control proposed for three months later or in case of worsening.

Introduction

The diagnosis of the several types of aortic arch diverticulum is important due to the difficulty in differing it from aneurysms and other mediastinal masses¹. Clinical and image diagnosis focuses on the reduction of mortality associated with rupture or compression of mediastinal structures.

Kommerell diverticulum is a rare condition that occurs more frequently in the left aortic arch (LAA) and anomalous origin of the right subclavian artery (RSA) (0.5-2.0%), but not very common in the right aortic arch (RAA) and anomalous origin of left subclavian artery (LSA) (0.05-0.1%)^{2,3}. The cause of anomalous origin of subclavian artery may be the abnormal regression of the fourth primitive aortic arch during embryonic development. The fourth left aortic arch persists as an aortic arch, while the fourth right aortic arch remains as a right subclavian artery and innominate artery^{3,4}.

Image exams, particularly magnetic resonance angiography (MRA) with 3D-GE TOF technique during administration of gadolinium – and the possibility of multiple-projection reconstructions (MPR) – play a fundamental role in the diagnosis and preoperative program due to the complexity of the surgical treatment^{2,3,5,6}.

The objective of this paper is to report the aspects of magnetic resonance angiography in a patient with Kommerell

diverticulum in the right aortic arch, with aberrant left subclavian artery.

Case report

Male, 65-year-old patient who referred having regurgitation (reflux) and retrosternal burning pain in the middle of the night. The patient was submitted to endoscopy and peptic ulcer treatment, but the symptoms lingered even after adequate treatment. In the endoscopic approach, an extrinsic steepening of the upper esophagus was identified and, thus, a thoracic radiography was indicated for the assessment of a mediastinal mass.

After the exam, that showed mediastinal enlargement of apparent vascular origin, the patient was referred to the image diagnostic service of Hospital das Clínicas de Niterói, Rio de Janeiro, Brazil, for magnetic resonance angiography procedure (Figures 1 and 2).

The 3D-GE TOF magnetic resonance angiography with gadolinium can identify the aortic arch at the right side with the diverticular colon and the origin of left subclavian artery. The trachea and the esophagus were in anterior position in relation to the vascular structures, as easily identified by the exam.

The study with 3D reconstruction of the thoracic aorta showed a better aortic arch at the right side, as well as aberrant left subclavian artery with origin in the Kommerell diverticulum.

The patient presented clinical improvements after taking prokinetics medication and proton pump inhibitors, as clinical follow-up and a new image exam was indicated for three months later or in case of clinical worsening.

Discussion

There are basically three types of aortic arch diverticulum: the diverticulum at left aortic arch with aberrant right subclavian artery; the diverticulum at right aortic arch with aberrant left subclavian artery; and, finally, the aortic diverticulum at aortic-ductal junction^{4,7-9}.

The first type of diverticulum was described by Kommerell⁵, in 1936, in a publication on a case of aberrant right subclavian artery, the most common anomaly of the subclavian artery, which originates in the thoracic aorta with arch at the left associated with right remnant dorsal aorta. The latter is similar to the diverticulum from which the correspondent subclavian artery originates. The Kommerell diverticulum, which corresponds to a conical dilatation in the origin of the aberrant vase, is also known as "lusoria diverticulum", "remnant diverticulum" or "lusoria root"⁵.

Key Words

Aorta, Thoracic; Subclavian Artery / abnormalities; Kommerell Diverticulum.

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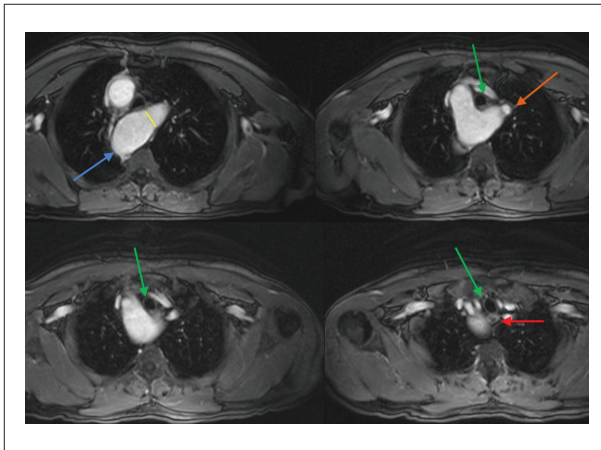


Figure 1 – Aortic arch at the right side (blue arrow) with rectal diverticulum (yellow line) from where the left subclavian artery originates (orange arrow). Observe the trachea (green arrow) and the esophagus (red arrow) anterior to vascular structures.

The second diverticulum type is described in dextroposed aortic arch as a rare congenital alteration (present in 0.05 to 0.1% of radiologic series^{6,7} and 0.04 to 0.1% of necropsy series^{7,9,10}). It may be associated with the aberrant left subclavian artery^{3,5}, which may be localized behind the esophagus (80%), precisely between the trachea and the esophagus (15%) or behind the trachea (5%)^{5,7,8}. The right aortic arch results from an anomalous primitive organogenesis. In general, between the fourth and fifth weeks of embryonic life, the blood leaves the heart through a single vase, the arterial trunk, which is divided into two branches: right and left ventral aorta. They communicate with the dorsal artery, respectively, through six brachial vases, known as aortic arches. The first three arches, together with their ventral and dorsal aortic connections, form the carotid system. A segment of the ventral aorta, the fourth right arch and a portion of the right dorsal aorta originate the right subclavian artery and the brachiocephalic trunk (innominate artery). The fourth left arch persists and becomes the adult aortic arch that unites with the seventh dorsal intersegmental artery to form the left subclavian artery. The fifth arches are reabsorbed, while the sixth arches form the pulmonary and arterial trunks. If the fourth left arch disappears and the right arch persists, a right aortic arch will develop. Generally, it is associated with the left dorsal aorta involution and with the right dorsal aorta persistence, resulting in a descending thoracic aorta placed in the right hemithorax. If the right dorsal aorta disappears, the dextroposed aortic arch places behind the esophagus to unite to the left dorsal aorta, while the descending thoracic aorta will unite to the left one^{3,8}. If both arches persist, they will form a double aortic arch or a vascular ring that encircles the trachea and the esophagus^{9,10}.

Among the proposed classifications based on anatomical distribution of the involved structures, the most widely used is Edwards classification^{4,6}. Three types of right aortic arches have been described¹: type I, when the big arteries branches form a mirror-image; type II, with artery segment enclosing area; and type III, when the isolated left subclavian artery communicates with the pulmonary area through the artery duct.

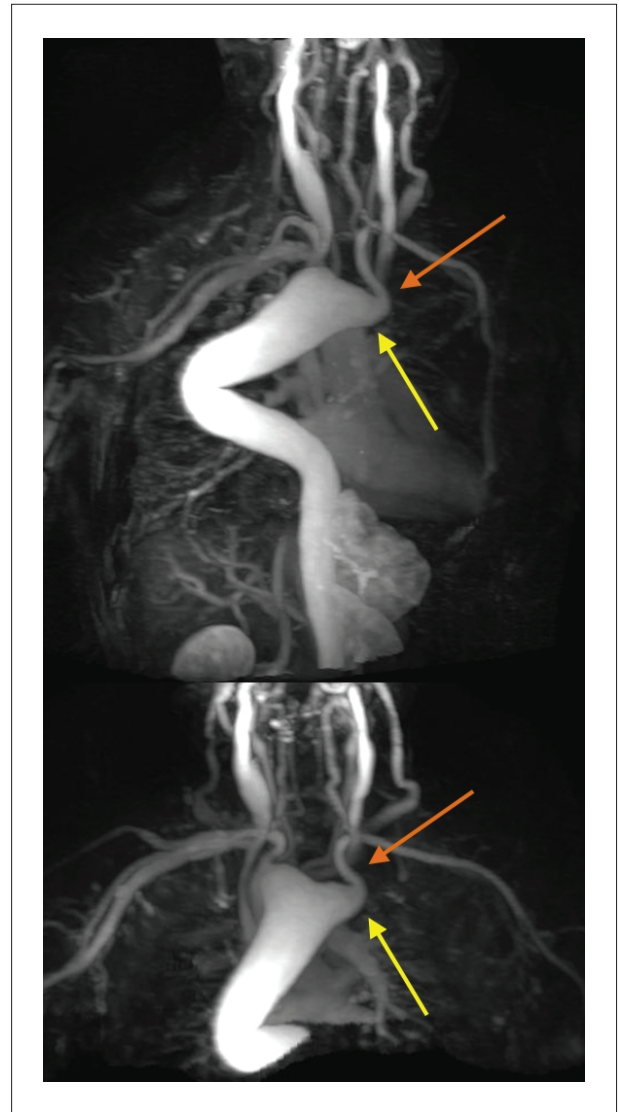


Figure 2 – Magnetic resonance angiography with 3D reconstructions of the thoracic aorta, making evident an arch at the right side and aberrant left subclavian artery (red arrow) that originates in Kommerell diverticulum (yellow arrow).

In the present case report, the aortic arch dextroposition was associated with the aberrant left subclavian artery (Edwards' type II), which in 5 to 10% of the cases may be related to congenital cardiopathies, including Tetralogy of Fallot, pulmonary stenosis with ventricular septal defect, tricuspid atresia and ductus arteriosus². The right aortic arch is frequently asymptomatic. In adults, symptoms may originate from atherosclerotic disease of anomalous vases, such as aneurysm dissection or dilatation with compression of adjacent structures, causing dysphagia (dysphagia lusoria), dyspnea, stridor, noisy breathing, cough, repetition pneumonia, obstructive emphysema or thoracic pain^{2,3}.

The third type is the aortic diverticulum in the aortic-ductal junction^{7,9,10}. This diverticulum shows a steepening throughout the internal portion that comprises the distal aortic

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isthmus till the left subclavian artery. It is also known as "ductal diverticulum", "ductal steepening" or "aortic diverticulum"⁹. The term "ductal diverticulum" may also be applied to a steepening in the left pulmonary artery, which represents a remnant of the arterial duct pulmonary orifice^{6,10}.

Final considerations

Magnetic resonance angiography may be useful as a preoperative image exam in cases of surgical planning or when images are needed for ambulatory follow-up care.

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