Aberrant right subclavian artery and Kommerell's diverticulum associated with aortic dissection and vascular rupture.

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Learning objectives

The aberrant right subclavian artery is the most common embryological anomaly of the aortic arch and more than half of cases are associated with a Kommerell's diverticulum. These associated malformations can cause distal embolization, compression of adjacent structures, vascular rupture and dissection. New imaging modalities, such as the case of cardiovascular multisliced computed tomography allow us to better assess the complex and unusual vascular diseases. Our aim is to review the embryology, anatomy, and imaging diagnostic of this disease, focusing on what the radiologist must know.

Background

A right-sided aortic arch is the result of an abnormal organogenesis of primitive aortic arches. Between the fourth and fifth weeks of embryonic life, blood leaves the heart by a single vessel, the truncus arteriosus, which divides into two branches, the ventral aortae. These are connected with the paired dorsal aortae by six branchial vessels, called aortic arches. Segments of the first three arches, together with their dorsal and ventral aortic connections, form the carotid system. A segment of the right ventral aorta, the right fourth arch, and a portion of the right dorsal aorta develop into the right subclavian artery and the innominate artery. The left fourth arch persists as the adult aortic arch, and with the anlagen of the seventh dorsal intersegmental artery it forms the left subclavian artery. The fifth arches are both resorbed, and the sixth arches form the pulmonary artery and the truncus arteriosus. The right-sided aortic arch results from persistence of the right fourth aortic arch and involution of the left. The right arch passes over the right main stem bronchus to the right of the trachea and esophagus. It is usually associated with involution of the left dorsal aorta and persistence of the right, causing the descending thoracic aorta to be located in the right hemithorax. If, instead, the right dorsal aorta disappears, the right-sided arch passes behind the esophagus to join the left dorsal aorta and the thoracic aorta descends in the left chest.

An aberrant left subclavian artery arising either as the last branch of the right-sided aortic arch or from an aortic diverticulum, called Kommerell's diverticulum, that is a remnant of the left dorsal aortic arch. This occurs as a result of the reabsorption of the left fourth aortic arch proximal to the origin of the left subclavian artery. Congenital heart anomalies are present in only 5% to 10%. Because of the atherosclerotic changes that occur in the arterial wall during life, in adults it is generally not possible to distinguish a true diverticulum (Kommerell's diverticulum, an embryonic remnant) from an acquired aneurysm of the origin of the aberrant subclavian artery.

Right-sided aortic arch may be asymptomatic. In infancy, symptoms are related to congenital heart anomalies or to compression of mediastinal structures such as the
trachea or the esophagus. In adulthood, symptoms are more often the result of early atherosclerotic changes of the anomalous vessels, dissection, or aneurysmal dilatation with compression of surrounding structures causing dysphagia (*dysphagia lusoria*), dyspnea, stridor, wheezing, cough, choking spells, recurrent pneumonia, obstructive emphysema, or chest pain.

Indirect evidence of vascular anomalies of the aortic arch and its branches may be obtained with barium swallow, esophagogastrosopy, and bronchoscopy showing indentation of the esophagus or compression of the upper airways. Diagnostic techniques include computerized axial tomography, angiography, and magnetic resonance angiography.

Definition of the anatomy of the aortic arch vessels may prove difficult even with angiography; occasionally, an incorrect diagnosis of aortic dissection has been made. Multiple projections are often required, and some authors suggest a left transaxillary route for angiography to reduce problems with interpretation of findings. However, the combination of thin-section computerized axial tomography and angiography is usually able to define vascular anatomy.

An aneurysm involving the distal arch, the origin of the left subclavian artery, and the thoracic aorta may be repaired through a right thoracotomy. A left subclavian-to-carotid transposition before the thoracic repair allows a practical approach to reconstruction of the subclavian artery. Careful preoperative imaging and consideration of the individual anatomy in surgical planning are essential to a successful outcome.

**Imaging findings OR Procedure details**

We report a dissection of the distal arch and descending thoracic aorta in a patient with right-sided arch Fig. 1 on page 5 Fig. 2 on page 5 Fig. 3 on page 6. In our patient, the left subclavian artery originated at the junction between the distal arch and the descending thoracic aorta located in the right chest and was aneurysmal (Kommerell's diverticulum) Fig. 4 on page 7 Fig. 5 on page 8 Fig. 6 on page 9.

The patient was a 50-year-old obese male with hypertension and obesity, no malformation was known previously and took no medication.

The dissecion involved too the abdominal aorta, left iliac artery, celiac artery, hepatic artery, splenic artery, left renal artery and inferior mesenteric artery.

No perfussion defects were observed at the abdominal organs Fig. 7 on page 10 Fig. 8 on page 11 Fig. 9 on page 12.

The patient was treated conservatively with good evolution, but died suddenly three months later (no necropsy was performed).
Images for this section:

Fig. 1: Undamaged ascending aorta
Fig. 2: Coronal view of descending aorta and iliac division
Fig. 3: Sagittal view of descending aorta
Fig. 4: Kommerell's Diverticulum
Fig. 6: Kommerell's Diverticulum. Volume rendering
Fig. 7: Thoracic aorta
Fig. 8: Upper abdominal aorta
Fig. 9: Lower abdominal aorta
Conclusion

Acquired cardiovascular diseases can modify the clinic manifestations of a vascular congenital anomaly. Our case is an example of how, with the use of advanced medical technology, we can study with detail those complex pathologies. As times goes by, frequency of interactions between congenital and acquired conditions may increase because of the growing of the prevalence of vascular diseases in the population. The radiologist must know this and include this possible association in the diagnostic evaluation of the patients.

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