

Type II right aortic arch with aberrant left subclavian artery and aortic diverticulum. A case report

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Section: Cardiovascular

Area of Interest: Cardiovascular system Thorax Arteries
/ Aorta Computer applications Vascular

Procedure: Contrast agent-intravenous

Procedure: Diagnostic procedure

Procedure: Computer Applications-3D

Imaging Technique: CT-Angiography

Imaging Technique: Image manipulation /
Reconstruction

Special Focus: Congenital Case Type: Clinical Cases

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Patient: 77 years, male

Clinical History:

A patient with embolic infarction was referred for CTA, due to a suspected stenosis of the ICA.

Regarding the found anomaly, the patient was asymptomatic and no further treatment was required. Due to the symptomatic stenosis of the ICA, the patient was referred for carotid endarterectomy.

Imaging Findings:

A stenosis of the C1 segment of the right internal carotid artery was detected (70% according to NASCET). As an incidental finding, the arch of the aorta was located to the right of the trachea, crossing over the right main bronchus (Figs' 1, 3). The first branch from the aortic arch was the left carotid artery; the second branch was the right carotid artery; and the third branch was the right subclavian artery. The left subclavian artery was the most dorsal branch (Fig. 1), originating from an aneurysmal dilatation (Figs' 2a & 3b) and measuring 2.8 x 2.4 cm (aortic diverticulum) dorsal to the oesophagus (Fig. 2b) (aberrant left subclavian artery). The descending aorta then passed to the right of the vertebral column and later it shifted to the midline to enter the abdomen through the aortic hiatus in the diaphragm (Fig. 2a).

Discussion:

When the embryo reached approximately 30 mm, the ventral aortic sac and the paired dorsal aortae become connected to each other through 6 pairs of vessel channels. The fourth pair begins to appear at about the end of the fourth week. At the left side, it will continue to grow forming the adult aortic arch. On the right side it partially forms the right subclavian artery and its caudal part will regress [1].

If the right-sided fourth channel, instead of the left, continues to grow with regression of the caudal part of the left fourth channel, then a right-sided aortic arch will form [1].

It is classified in 3 categories:

Type I with mirror-image branching; type II with aberrant left subclavian artery, and type III, with isolation of the left subclavian [2].

Right-sided aortic arch is found in approximately 0.1% [2].

The condition is usually asymptomatic and discovered incidentally. The most commonly described symptom is

dysphagia, or dysphagia lusoria, which comes from the Greek “*lusus naturae*”, meaning freak of nature, and was termed in 1761 by the English surgeon David Bayford [3].

Association of this condition with congenital heart diseases will lead to earlier detection of the anomaly. The combination of right aortic arch and tetralogy of Fallot is called Corvisart disease, which was named in 1818 by Bonaparte's primary physician Jean Corvisart [2, 4].

On chest X-ray the usual aortic knob will be absent and a right-sided paratracheal shadow may be seen. Indentation on the posterior surface of the oesophagus will be noticed on barium swallow. CT will show the anomaly clearly. In some cases, a dilatation of the origin of the left subclavian artery will be seen. This represents a remnant of the distal primitive left arch [5]. It is called “Kommerell's diverticulum” after the German radiologist Burckhard Kommerell who, in 1936, first described an aortic diverticulum at the origin of an aberrant right subclavian artery, in a patient with presumed gastric cancer during barium studies [5, 6].

MRA is an alternative investigation with the advantage of avoiding radiation.

The condition has a good prognosis if it is not associated with further congenital anomalies [7]. Incidentally discovered cases require no treatment.

Teaching point:

The presence of right-sided aortic arch may explain symptoms such as dysphagia; or it may alert the attention to a possible congenital heart disease. Precise anatomical description of the vascular anomaly is necessary if a surgery required.

Differential Diagnosis List: Type II right aortic arch with aortic diverticulum., Double aortic arch, Kommerell's diverticulum at the origin of an aberrant right subclavian artery in a left-sided aortic arch, Aortic diverticulum at the aortoductal junction, Aneurysms and traumatic pseudoaneurysms of the aorta, Other mediastinal masses (chest x-ray)

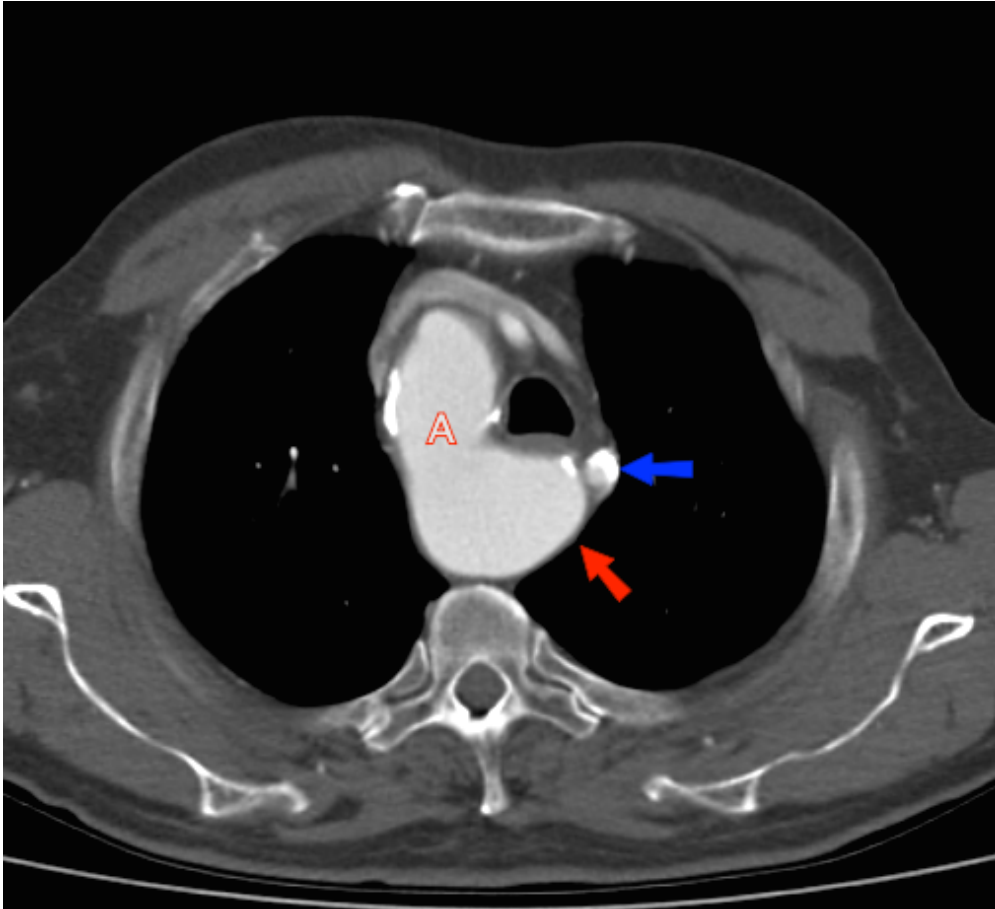
Final Diagnosis: Type II right aortic arch with aortic diverticulum.

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Figure 1

a



Description: Axial CT showing the aortic arch (A) to the right side with a diverticular dilatation (red arrow) dorsal to the trachea. The last branch from the aortic arch is the left subclavian artery (blue arrow). **Origin:** Hakim A, Department of Radiology, KKH Lörrach. Germany

Figure 2

a



Description: Coronal oblique reformation. Fig a. shows the aortic diverticulum (red arrow), from which the aberrant subclavian artery (blue arrow) originated. **Origin:** Hakim A, Department of Radiology, KKH Lörrach, Germany

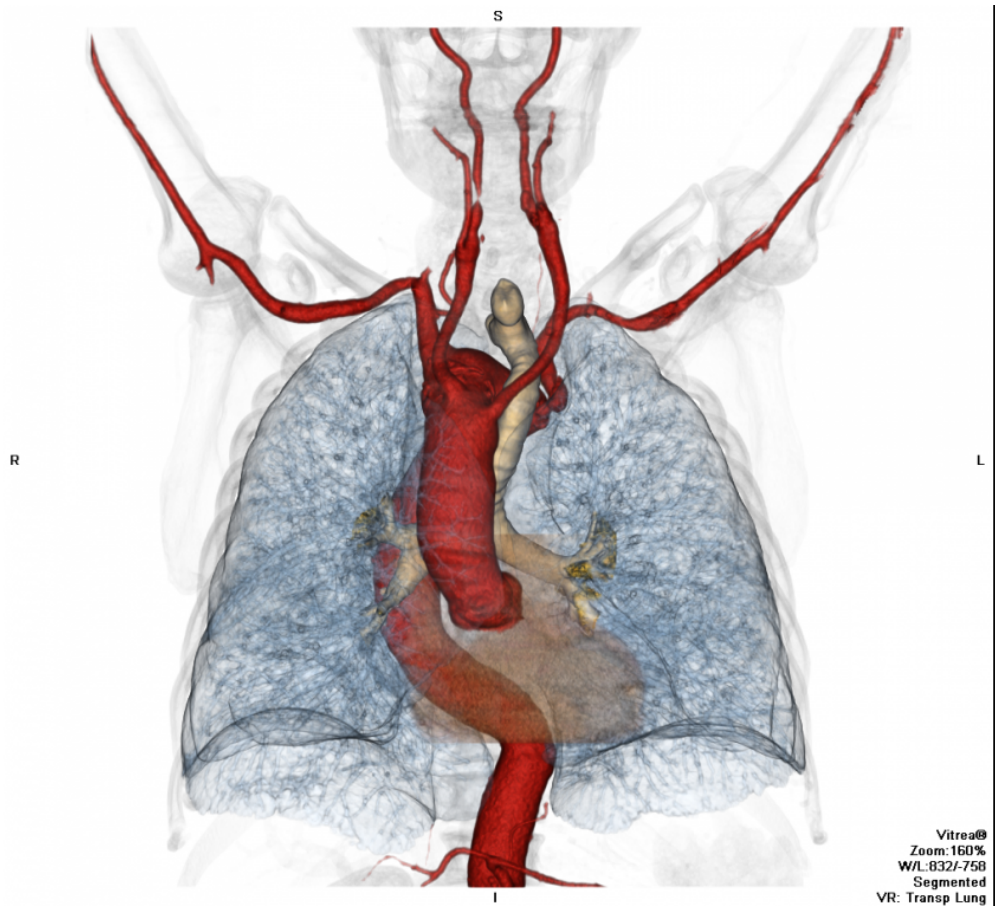
b



Description: Sagittal curved reformation shows the relation between the esophagus (white arrows) and the aortic diverticulum **Origin:** Hakim A, Department of Radiology, KKH Lörrach, Germany

Figure 3

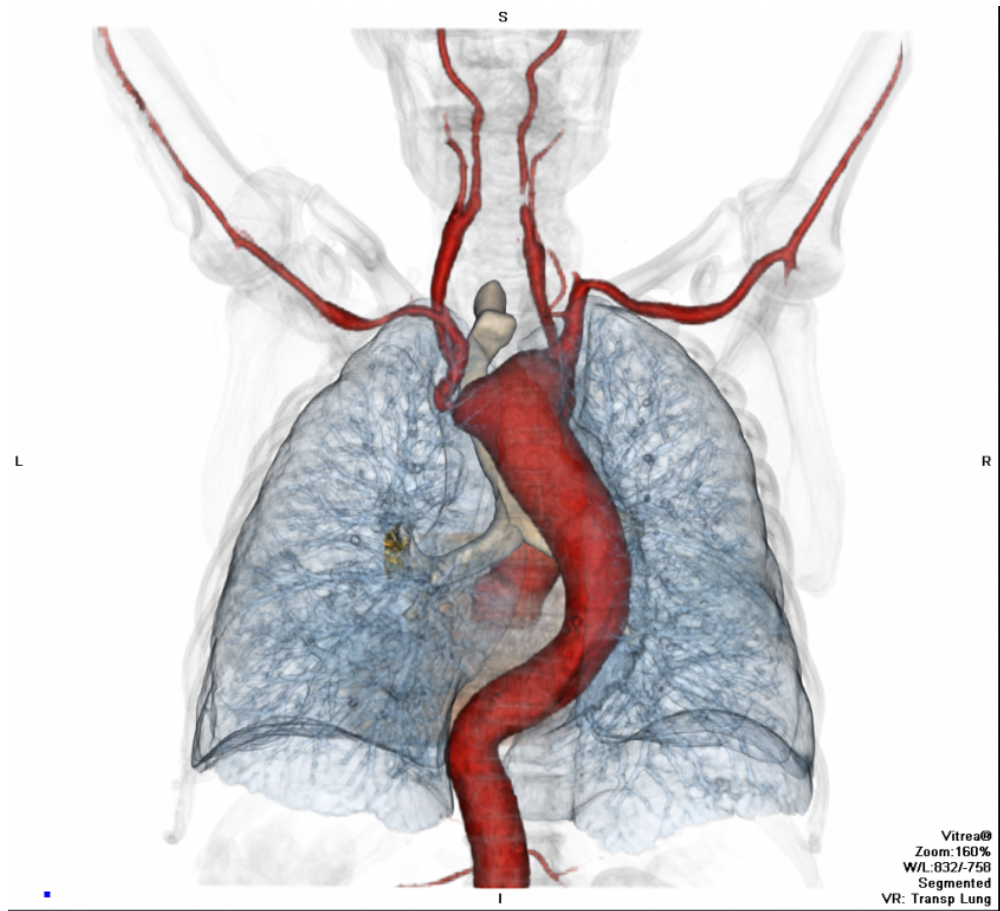
a



Description: Post processing of the CT in frontal view, shows a right-sided aorta crossing over the right main pulmonary bronchus.

Note: stenosis of the right ICA. **Origin:** Hakim A, Department of Radiology, KKH Lörrach, Germany

b



Description: Post processing of the CT in dorsal view. The distal portion of the aortic arch shows a diverticular dilatation, from which the left aberrant subclavian artery originated. **Origin:** Hakim A, Department of Radiology, KKH Lörrach, Germany