

Right-sided aortic arch and an aberrant left subclavian artery: a case report

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SUMMARY

A right-sided aortic arch is a rare congenital defect of the aorta. A 56-year-old Caucasian male patient was hospitalized with coronary artery disease (stable angina CCS II, postinfarction atherosclerosis), stage II hypertension, impaired glucose tolerance and congestive heart failure NYHA class II. Contrast-enhanced ECG-gated multi-slice computed tomography was performed on Siemens SOMATOM Force Germany and revealed right-sided aortic arch, aberrant left subclavian artery with Kommerell's diverticulum and kinking of the thoracic aorta. This case is an example of the great advantages of knowing human anatomy and embryology in clinical practice. Modern diagnostic modalities give an accurate information on congenital variants and anomalies of the aortic arch and its branch that is vital for vascular surgery in the thorax, head and neck region.

Key words: Aorta – Aberrant left subclavian artery – Congenital heart disease – Kommerell's

diverticulum – Right-sided aortic arch – Multi-slice computed tomography

INTRODUCTION

A left- or right-sided aortic arch (RSAA) refers to the position of the aortic arch in relation to the trachea (Miranda et al., 2014). The normal left aortic arch lies on the left of the trachea and courses over the left main bronchus. A RSAA refers to an aortic arch that courses and descends to the right side of the trachea.

A right-sided aortic arch is a rare congenital defect of the aorta and occurs in 0.05% to 0.1% of radiology series and in 0.04%-0.1% of autopsy series (Arazinska et al., 2017; Priya et al., 2018; Popieluszko et al., 2018). The real incidence of RSAA in the general population is unknown, but a rate of 1/1000 has been suggested by Achiron et al. (2002). The recent progress in perinatal medicine and imaging techniques had facilitated the prenatal diagnosis of this cardiovascular anomaly (Muraoka et al., 2018). RSAA was detected in 0.6% of all fetal

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cardiac examinations and to 5.0% of all cardiac abnormalities (Miranda et al., 2014). Abnormalities of aortic arch orientation are often associated with a variety of congenital heart defects (Tetralogy of Fallot and truncus arteriosus), as well as chromosomal abnormalities, such as DiGeorge syndrome (22q11 deletion) (Law and Mohan, 2019).

There are three main subtypes of RSAA according to Edwards' model: type I – RSAA with mirror image branching; type II – RSAA with aberrant left subclavian artery (ALSA); and type III – RSAA with isolated subclavian artery. The most common subtype is type II, when the right common artery arises as the first branch and is followed by the right subclavian artery and the left common carotid artery while the left subclavian artery is the last branch and takes a retroesophageal course to reach on the left side (García-Guereta et al., 2016; Priya et al., 2018). The aberrant left subclavian artery has a variable descending: posterior to the esophagus in 80% of cases, anterior to the trachea in 5% of cases and just between the trachea and esophagus in 15% of cases (Tong et al., 2015). Edwards' type II reveals cardiac anomalies only in 5-10% of cases, which are commonly diagnosed in newborns and young children and rarely become symptomatic in adults (Ebner et al., 2013). About half of the patients with right-sided aortic arch have an aberrant left subclavian artery, which may arise either directly from the aorta or from the Kommerell's diverticulum (Tong et al., 2015). The complete vascular ring is formed by the segment of the ascending aorta anteriorly, Kommerell's diverticulum posteriorly and the ligamentum arteriosum coursing on the left side of the trachea. Any symptoms, which result from an aberrant left subclavian artery, are associated with compression of the esophagus or trachea and are most likely to occur if its origin is dilated (Kakaria et al., 2008; Yang et al., 2009).

In this article, we report an asymptomatic patient with right-sided aortic arch in whom an aneurysm of the aberrant left subclavian artery was detected.

CASE REPORT

A 56-year-old Caucasian male patient was hospitalized in the Belarusian State Institution

«Republican Scientific and Practical Centre «Cardiology» in 2018 for typical angina combined with shortness of breath upon walking about 1000 m and climbing stairs to the 4th floor. He had a history of an acute myocardial infarction in 2017. The patient provided the informed consent regarding radiological images, clinical assessment and publication of patient's data.

He was diagnosed with chronic coronary artery disease (stable angina CCS II, postinfarction atherosclerosis) based on typical clinical presentation, hypertension stage III, impaired glucose tolerance and congestive heart failure NYHA class II. Contrast-enhanced ECG-gated multi-slice computed tomography, performed on Siemens SOMATOM Force (Germany), revealed:

The right-sided aortic arch (Fig. 1) crossing the midline at the level of the Th4-Th5 vertebral bodies, following a retroesophageal path and continuing to the descending part of the aorta, which was situated on the left side of the vertebral column.

Kommerell's diverticulum (Fig. 1), a bulbous formation at the point of origin of the aberrant left subclavian artery with a diameter of 36 mm.

Kinking of the thoracic aorta at the level of the arch (Fig. 1). The diameter of the aorta fell within the normal range: 26 mm at the level of the valve's annulus; 36 mm at the level of the aortic sinuses; 25 mm at the level of the sinotubular junction and proximal ascending aorta was measured at 27 mm. The diameter of the thoracic region of the descending part of the aorta was expanded to 49 mm.

The branches of the aortic arch – the right subclavian artery, the right and left common carotid arteries – did not show any stenosis. The pulmonary trunk was not expanded. Our patient did not have any cardiac and vascular congenital abnormalities associated with RAA and aberrant LSCA.

COMMENTS

The aberrant right subclavian artery accounts for 0.5-1.8% of the population as the most frequently encountered aortic arch anomaly

(Polguy et al., 2013; Naqvi et al., 2017), while right-sided aortic arch and an aberrant origin of the left subclavian artery with Kommerell's diverticulum is a rare congenital anatomical condition most often observed in adults (Silveira et al., 2019). In the current report, we presented a classic case of this malformation diagnosed accidentally in

which the aberrant left subclavian artery arises from the retroesophageal diverticulum located behind the esophagus. The complete vascular ring encircling of trachea and esophagus was not formed, and therefore there were no clinical signs of esophageal compression. The approach to asymptomatic patients with vascular ring

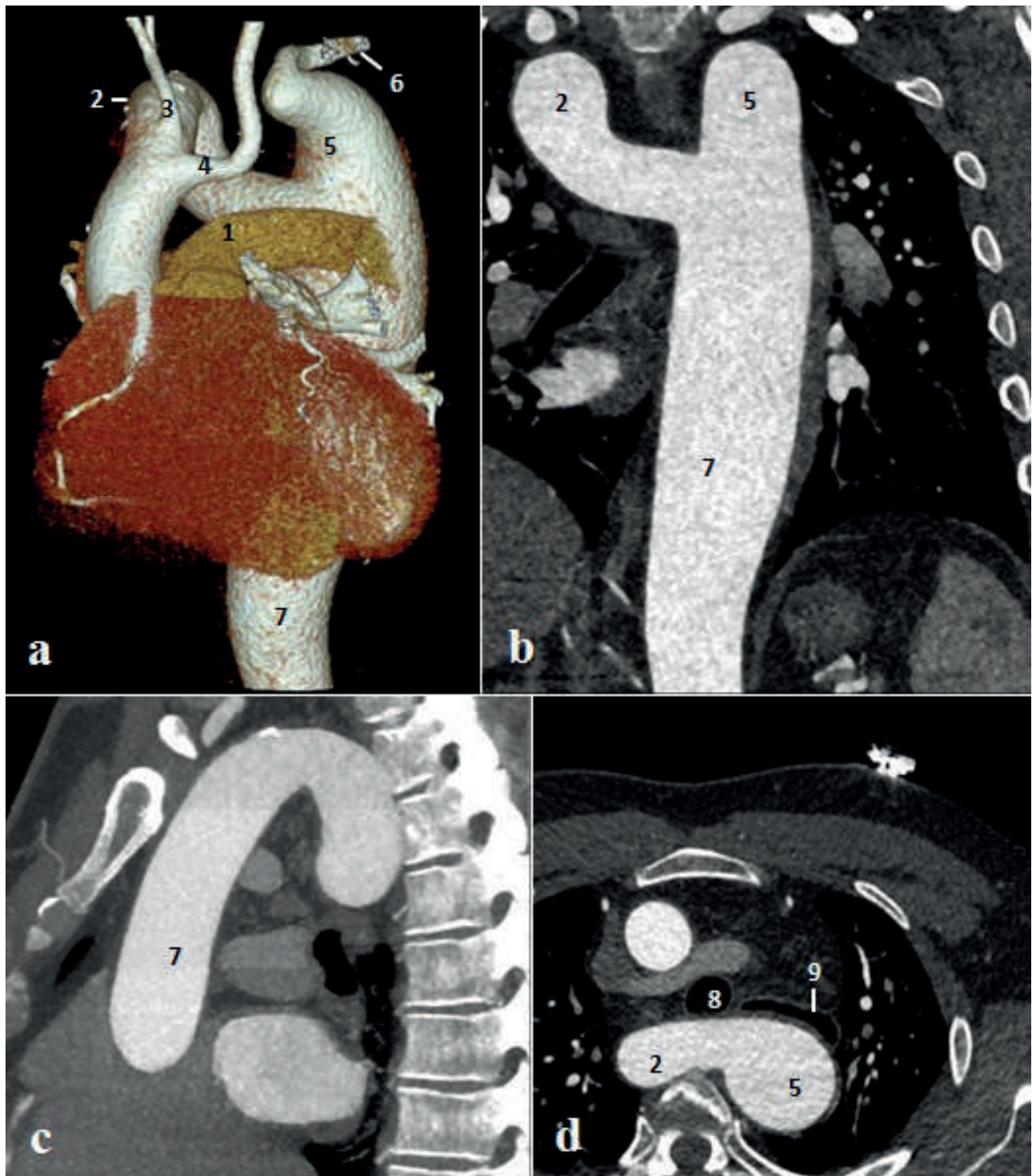


Fig. 1.- Right-sided aorta and its branches: **a** – CTA VRT image, front view; **b** – CTA MIP image, coronal section; **c** – CTA MIP image, sagittal section; **d** – CTA MIP image, axial section. 1. Pulmonary trunk. 2. Right-side aortic arch. 3. Brachiocephalic trunk. 4. Left common carotid artery. 5. Kommerell's diverticulum. 6. Aberrant left subclavian artery. 7. Descending thoracic aorta. 8. Trachea. 9. Esophagus.

(VR) is controversial because this condition is usually diagnosed during pregnancy and affected individuals can remain asymptomatic for a long period of time (García-Guereta et al., 2016). Most centers perform some type of postnatal confirmatory tests of the VR, particularly echocardiography, but due to the limitations of echocardiography magnetic resonance imaging has been proposed.

Aneurysmal diverticulum itself can cause the compression of the esophagus or may rupture spontaneously (Priya et al., 2018). The diverticulum is usually resected in symptomatic patients and followed by the aortopexy of the arch. Alternatively, after the resection of diverticulum, the left subclavian artery can be re-anastomosed to the left carotid artery.

The embryological origin of the various anomalies of the aortic arch is based on the theory of the hypothetic double aortic arch, which was described by Edwards (Yoo et al., 2003; Hsu et al., 2011). The ascending (ventral) aorta is divided into two aortic arches, which surround each side of the trachea and esophagus and connect to form the descending (dorsal) aorta. A common carotid artery and subclavian artery are derived from each aortic arch. Normally, the regression occurs between the origin of right subclavian artery and descending aorta in the right ascending aorta. Thus, the normal aortic arch is formed. If the interruption presents between the left carotid artery and the left subclavian artery in the left aortic arch, it will form a right-sided aortic arch with the aberrant left subclavian artery. There can be a dilated segment at the proximal part of the ALSA. The proximal part of the aberrant artery carries the blood flow from the ductus arteriosus into the descending aorta; and, therefore, the proximal part of the aberrant artery is as wide as the ductus and descending aorta (Hsu et al., 2011).

This case is a clear example of the helpfulness of the human anatomy and embryology knowledge in clinical practice. Accurate information about congenital variants and anomalies of the aortic arch and its branch obtained with modern diagnostic imaging modalities is vital for vascular surgery in the thorax, head and neck region.

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