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ANATOMIC VARIATIONS

Coexistence an aberrant right subclavian artery with other congenital anomalies: case report and review of the literature

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Abstract
Aberrant right subclavian artery is the most common aortic arch anomaly that frequently occurs in coexistence with other congenital cardiovascular anomalies. A 32-year-old male patient was hospitalized with ventricular septal defect, chronic heart failure NYHA class III, pulmonary arterial hypertension. Contrast-enhanced multislice computed tomography revealed membranous ventricular septal defect, persistent left superior vena cava, bicuspid aortic valve and aberrant right subclavian artery. Aberrant right subclavian artery was clinically silent and discovered accidentally. The patient underwent heart–lung transplantation due to pronounced, irreversible pulmonary hypertension. This article reports a rare coexistence of aberrant right subclavian artery with other congenital anomalies of the heart and great vessels in living men.

Keywords Aberrant right subclavian artery · Bicuspid aortic valve · Persistent left superior vena cava · Ventricular septal defect · Congenital heart disease · Pulmonary arterial hypertension · Arteria lusoria

Introduction

Aberrant (anomalous) origin of the right subclavian artery from the aorta distal to the normally positioned left subclavian artery is one of the more frequent congenital anomalies in subjects with left aortic arch. In 1735 Hunauld first described this anatomic rarity from autopsy studies.

The frequency of aberrant right subclavian artery (ARSA) varies throughout the world [9]. In Europe, depending on the country, it has been found in 0.11% (Great Britain), 0.16% (Greece), 0.3% (France), or 0.36% (the Netherlands) of the population. Studies have also been performed on other continents: Asia, 0.1–0.2% of cases (respectively, China and Japan), North America, 0.5% of cases (United State), Australia and Oceania, 0.8% of cases (New Zealand).

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It should be noted that ARSA is, in general, an asymptomatic benign finding; but around 10% of subjects may complain of tracheo-oesophageal symptoms which usually present in the fourth or fifth decades [16]. A right retrooesophageal subclavian artery can cause dysphagia, dyspnea or chronic cough due to compressive mechanisms with contiguous organs. Due to the fact that the aberrant right subclavian artery can cause dysphagia, it is known clinically as "arteria lusoria" (AL).

This anomaly is attributed to involution of the 4th aortic arch during the 6th to 8th week of gestation. As a result, the 7th intersegmental artery remains attached to the descending aorta [9]. The root of the ARSA is formed by the persisting right aortic arch and as a result a diverticulum at the proximal descending aorta can be found. Once such primordial altered vascular arrangement has been completed, instead of being the normal first branch of the left-sided aortic arch (with the right common carotid as the brachiocephalic trunk), ARSA arises on its own as the fourth branch from the posterolateral portion of the distal arch (after the left subclavian artery) [16].

This article reports a rare coexistence of ARSA with other congenital anomalies of the heart and great vessels found in a male before cardiopulmonary transplantation.

Case report

A 32-year-old male patient was hospitalized to Belarusian State Institution «Republican Scientific and Practical Centre «Cardiology» in 2016 for breathlessness and decreased exercise tolerance. Signed informed consent was obtained for the patient both for the treatment and for using his personal data for publication.

He was diagnosed with ventricular septal defect; chronic heart failure NYHA class III; pulmonary arterial hypertension. Dual source 384-slice prospective ECG-gated chest CT angiography, performed on Siemens SOMATOM Force (Germany), revealed:

- Persistent left superior vena cava (PLSVC) with a diameter from 7 to 10 mm started from the left brachiocephalic vein and ended at great cardiac vein (Fig. 1a, b). Coronary sinus ostial atresia was detected (Fig. 1c) with drainage of the posterior interventricular vein into the great cardiac vein and PLSVC.
- Membranous ventricular septal defect (Fig. 1d) with a size of 24 mm.
- Bicuspid aortic valve type 1 according to Schaefer et al. (right-left-coronary-cusp fusion pattern) with calcification in the raphe of the conjoined coronary cusps (Fig. 1e). Ascending aorta diameter was 41 mm.
- Aberrant right subclavian artery (Fig. 2). It started from the left-sided aortic arch distal to the left subclavian artery at the level of the third intercostal space along the left circumferential line. Then the artery was directed up and to the right in the gap between the spine and esophagus, located at the level from the upper endplate of the fourth thoracic vertebra to the lower endplate of the body

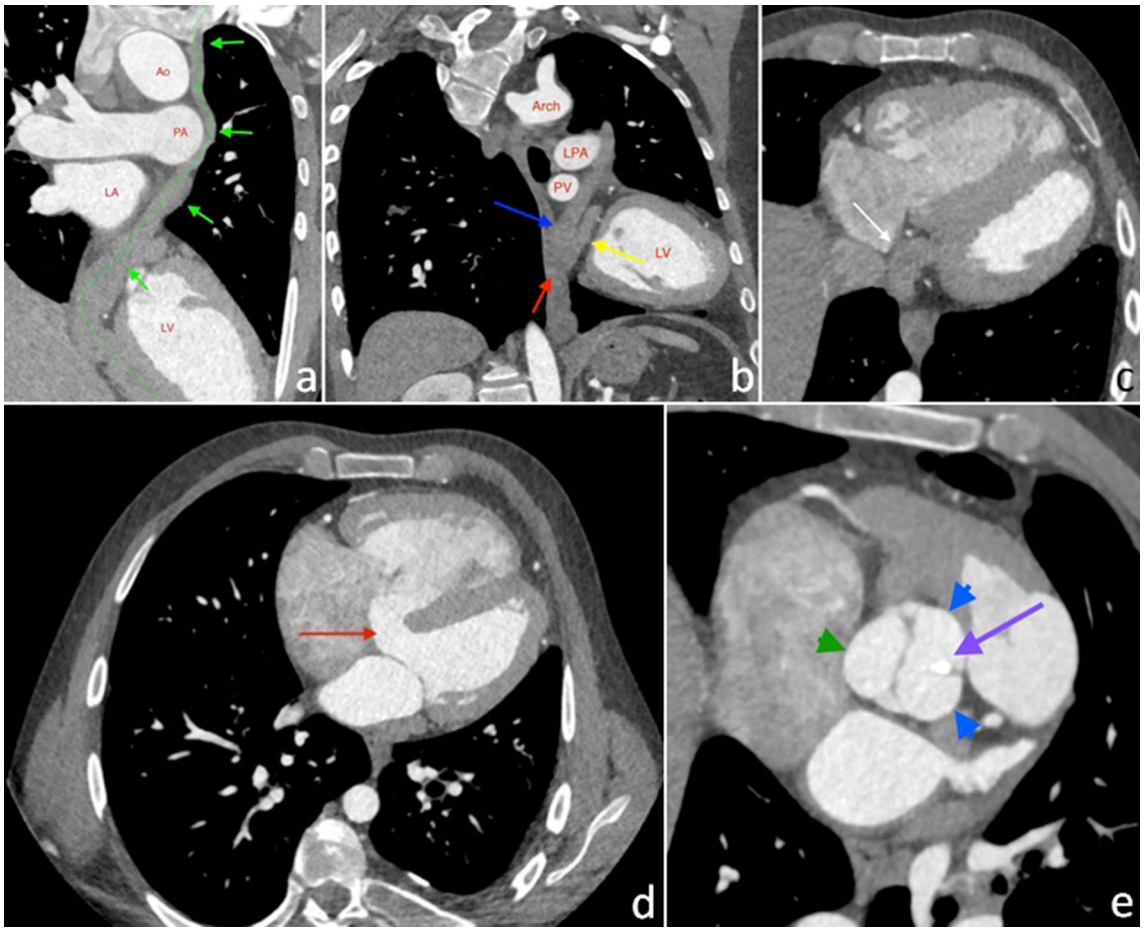

Fig. 1 Congenital cardiovascular malformations revealed by Contrast-enhanced ECG-gated multislice computed tomography: **a** curved CTA MPR images direction of the persistent left-sided superior vena cava (green arrows); **b** persistent left-sided SVC (blue arrow) connects to the vena cordis magna (yellow arrow), the junction is marked with a red arrow; **c** atresia of the coronary sinus ostium (white arrow); **d** ventricular septal defect (red arrow); **e** oblique transverse CTA reformation images of aortic valve (diastole). Raphe is located at fusion of right and left coronary cusps. Note the raphe calcification (purple arrow). Blue arrowheads show right and left semilunar cusps and green arrowhead shows posterior semilunar cusp. *Ao* Aorta, *PA* main pulmonary artery, *LA* left atrium, *LV* left ventricle, *Arch* aortic arch, *LPA* left pulmonary artery, *PV* pulmonary vein. (Color figure online)

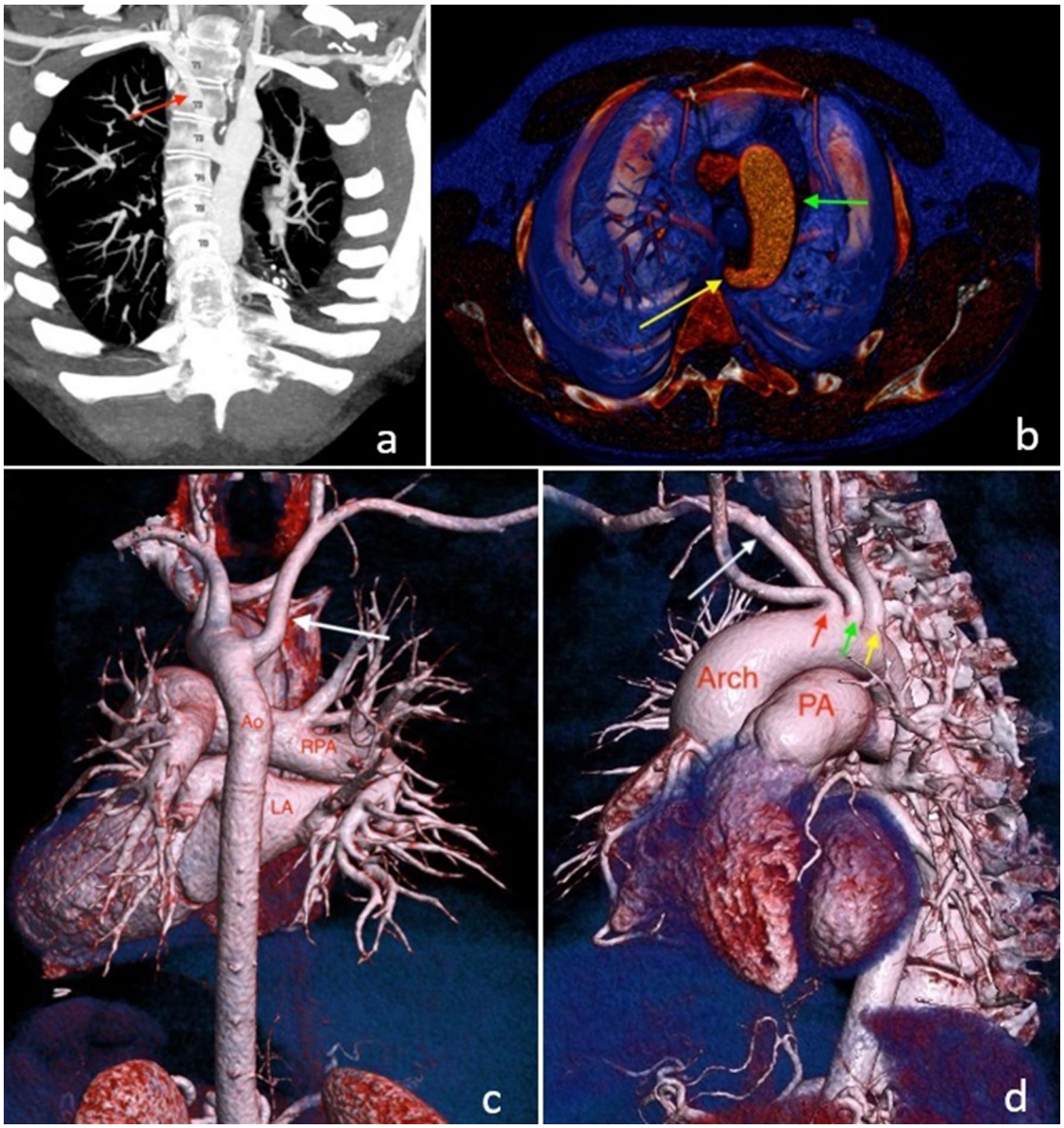

Fig. 2 Topography of aberrant right subclavian artery (ARSA): **a** CTA MIP image, coronal section; ARSA (red arrow); **b** CTA VRT image, inferior view; aortic arch (green arrow) and ARSA (yellow arrow); **c** CTA VRT image, lateroposterior view; ARSA (white arrow); **d** CTA VRT image, lateroanterior view; ARSA (white arrow); right common carotid artery (red arrow); left common carotid artery (green arrow), left subclavian artery (yellow arrow). *Ao* Aorta, *PA* main pulmonary artery, *Arch* aortic arch, *RPA* right pulmonary artery. (Color figure online)

of the first thoracic vertebra. It passed retro-esophageally at the level of the intervertebral disc between Th3/Th4. The diameter of the vessel at its orifice was 12 × 13 mm, in the second and third segments—6 × 7 mm. The patient had no clinical symptoms of compression of neighboring anatomical structures by ARSA. Pathology from other branches of the arch of the aorta was not revealed.

In addition, the patient showed signs of pulmonary arterial hypertension: widening of the right ventricular cavity, right ventricular myocardial hypertrophy up to 12 mm, enlargement of the pulmonary trunk to 37 mm, expansion of bronchial collateral circulation.

The patient underwent heart–lung transplantation due to pronounced, irreversible pulmonary hypertension. Such operation was carried out for the first time in the Republic of Belarus.

Discussion

The present patient was similar to type G according to the Adachi-Williams-classification of aortic arch structural configurations [5] where a left-sided aortic arch has four branches including an ARSA. The right subclavian artery arises from the distal portion of the aortic arch as its last branch. The other main branches (common right and left carotid arteries and the left subclavian artery) follow they normal trend.

Different courses of the ARSA relative to neighboring structures have been described. Aberrant right subclavian artery courses to the right behind the oesophagus in majority of the cases (80%), between the oesophagus and trachea in 15%, and anterior to the trachea or main stem bronchus in 5% [14]. Thus, our case presented typical ARSA course.

In the presented case aberrant right subclavian artery was clinically silent and discovered accidentally. According to Polguj et al. [8] the most commonly reported symptoms related to compression of adjacent structures by aberrant right subclavian artery were dysphagia (71.2%), dyspnea (18.7%), retrosternal pain (17.0%), cough (7.6%), and weight loss greater than 10 kg over a 6-month period (5.9%). It is known that dysphagia generally develops in older patients due to increased rigidity of the oesophagus itself or the vessel wall, elongation of the aorta, or formation of an aneurysm. Krupiński et al. [3] found significantly higher aberrant right subclavian artery lumen area at the level of oesophagus and lower distance between aberrant right subclavian artery and trachea than in non-dysphagia patients. Those measurements strongly support the theory of an increased lumen area of an aberrant right subclavian artery at the level of oesophagus leading to dysphagia lusoria.

Arteria lusoria is usually seen together with other anatomical variations, such as bicarotid trunk, Kommerell diverticulum, thyroidea ima artery, bilateral abnormal origin of the internal mammary artery, non-recurrent laryngeal nerve, and right-sided aortic arch [4–6]. But Pelizzo et al. [7] presented a case in which ARSA was not coexisted with the non-recurrent right ILN. They believe that this fact disproves the theory of the immutable association of these two anomalies. Furthermore, it can be coexisted with many cardiac anomalies (aortic coarctation, interrupted AA, tetralogy of Fallot, truncus arteriosus, transposition of the great arteries, and ventricular and atrial septal defects), genetic disorders such as Downs', Edwards, and DiGeorge syndromes, aneurysms, and arterioesophageal fistula.

In the presented case, the aberrant right subclavian artery was detected in combination with bicuspid aortic valve, persistent left superior vena cava and ventricular septal defect. To our knowledge, this is the first case of such combination of congenital cardiovascular malformation in a living adult patient.

Prevalence of bicuspid aortic valve (BAV) in general population is 1–2%. In this study, BAV type 1 (with one raphe) was recognized. As stated by Szymczyk et al. [12] it was the most common type observed in 0,7% of 2053 patients with cardiovascular diseases. However, the coexistence of BAV and right ARSA has only been reported in single cases, mostly in the paediatric patients. Among patients who underwent chest computed tomography (CT) for various reasons Tyczyński et al. [14] found out 72 persons with either right or left aberrant subclavian artery. Among them 7 cases of BAV and ARSA coexistence were identified. Moreover, bicuspid aortic valve could be connected with other congenital heart defects such as ventricular septal defects [1]. In the presented case, a congenital ventricular septal defect was coexisted with pulmonary hypertension, which led to irreversible changes in the pulmonary vessels. This was an indication for cardiopulmonary transplantation.

Persistent left superior vena cava (PLSVC) is the most common congenital malformation of the thoracic venous return and is present in 0.3–0.5% of individuals in the general population with a normal heart, and 10–11% in individuals with congenital heart diseases [2], such as anomalous connections of the pulmonary veins, aortic coarctation, tetralogy of Fallot, transposition of the great vessels as well as dextroversion. The coexistence of the PLSVC and ARSA is rare. In the literature there were single cases of such combination of congenital developmental anomalies which have been described in a dysmorphic SGA fetus [13] and a 21-day-old neonate with hypoplastic left heart syndrome [15].

Conclusion

Despite the fact that ARSA is the most common aortic arch anomaly, it is an incidental radiological finding. Arteria lusoria occurs with slightly higher incidence in patients with other congenital cardiovascular anomalies [11]. It is suggested that arteria lusoria is a developmental defect rather than a normal variant. ARSA is most susceptible to iatrogenic injuries during surgical and radiological interventions [10]. Detailed preoperative evaluations by contrast-enhanced chest CT scans might prevent damage to ARSA.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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