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Authors: P. Ostrowski, M. Bonczar, W. Przybycień, I. Zamojska, B. Kołodziejczyk, J. Walocha, M. Koziej

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P. Ostrowski et al., An aberrant right subclavian artery

P. Ostrowski, M. Bonczar, W. Przybycień, I. Zamojska, B. Kołodziejczyk, J.Walocha, M. Koziej

Department of Anatomy, Jagiellonian University Medical College, Krakow, Poland

Address for correspondence: Mateusz Koziej, Department of Anatomy, Jagiellonian University Medical College, Mikołaja Kopernika 12, 33-332 Kraków, Poland, tel: +48 888 202 628, e-mail: mateusz.koziej@gmail.com

ABSTRACT

An aberrant right subclavian artery (ARSA), also called "Arteria lusoria", is described as a right subclavian artery with a retro-esophageal course that most frequently originates as the most distal branch of the aortic arch. The aim of the following study was to present and thoroughly describe a case of an ARSA, its course, branches, and relation to the surrounding anatomical structures and discuss the clinical significance and embryology of this variant. During routine dissection, a 63-year-old male cadaver with an abnormal variant of the right subclavian artery was found. The right subclavian artery branched off from the aortic arch as the most distal branch. Subsequently, it coursed posteriorly to the trachea and esophagus at the level of T2 and T3. Abnormalities in the branching pattern of the right subclavian artery were also discovered, such as the right vertebral artery originating from the right common carotid artery as its first branch. The present study presents a case of an ARSA, which is a rare anatomical variant of the branches of the aortic arch. The course and branching pattern of an aberrant subclavian artery is quite variable, and each variant can be associated with different possible complications. Furthermore, the ARSA is associated with other cardiovascular anomalies, such as the Kommerells diverticulum. Therefore, knowledge about the possible variations of this anomaly may be of great importance for physicians who encounter this variant in their practice.

Key words: subclavian artery; ARSA; arteria lusoria

INTRODUCTION

An aberrant right subclavian artery (ARSA), also called "Arteria lusoria" is the most common variation of the aortic arch [16, 29]. It is described as a right subclavian artery with a retro-esophageal course that most frequently originates as the most distal branch of the aortic arch [17]. Embryologically, a left aortic arch with an ARSA results from interruption of the right arch between the right common carotid and right subclavian arteries [23]. When the pharyngeal arches develop during the fourth and fifth weeks of embryological development, each pharyngeal arch obtains its own artery, or aortic arch, which arises from the aortic sac, and cranial nerve. The common carotids and the first part of the internal carotid arteries are formed by the third aortic arch. Furthermore, the fourth aortic arch persists on both sides and forms different structures on the right and left sides. The left fourth aortic arch forms part of the arch of the aorta, between the left common carotid and the left subclavian arteries. On the right side, it forms the proximal segment of the right subclavian artery. The distal part of the said artery is formed by a portion of the right dorsal aorta and the seventh intersegmental artery [27].

The prevalence of this vascular anomaly is said to be ranging from 0.16% to 4.4% in the general population, with a female predominance [16, 20]. The ARSA was first described by Hunauld in 1735 [9]. However, the term "dysphagia lusoria" was first mentioned by Bayford in 1787 in a study about a patient with a long history of dysphagia caused by an ARSA [3]. A scheme presenting ARSA can found in Figure 1.

Being aware of the possible anomalies of the aortic arch is important because they might be associated with vascular rings, heart diseases, and chromosomal abnormalities [7, 24]. Congenital variants such as the left circumflex aorta or the right-sided aortic arch are examples of anomalies that have been associated with the formation of vascular [7, 22]. Furthermore, the prevalence of the ARSA is said to be higher in patients with trisomy 21 (35%) [5].

Variations of the arterial system are frequently observed by medical professionals of many distinct specialties worldwide and oftentimes influence daily clinical practice in the form of treatment options [11, 15, 31]. Patients with an ARSA are usually asymptomatic. However, when symptoms are present, they usually consist of dysphagia, chest pain, hemoptysis, and hoarseness, among others [30]. Puri et al. described the symptoms, when present, as manifesting at two extremes of life [26]. In children, dysphagia and symptoms associated with tracheal compression might occur. However, respiratory symptoms associated with the ARSA rarely occur because the adult trachea is more rigid compared to the infant trachea which is more compressible.

The aim of the following study was to present and thoroughly describe a case of an ARSA, its course, branches, and relation to the surrounding anatomical structures. Furthermore, the clinical significance and embryology of this variant will be discussed.

CASE REPORT

During routine dissection, a 63-year-old male cadaver with an abnormal variant of the right subclavian artery was found. The right subclavian artery originated from the aortic arch as the most distal branch. Subsequently, it coursed posteriorly to the trachea and esophagus at the level of T2 and T3. At the level of the 1st intercostal space, a costocervical trunk branched off posteriorly. Afterward, a thyrocervical trunk originated superiorly to the first rib. Subsequently, the right subclavian artery exited the thorax via the superior thoracic aperture between the anterior and middle scalene muscles. The further course shows no exceptions from the currently adopted normal anatomy of the human body.

In the present case, the brachiocephalic trunk was absent, and the right common carotid artery originated directly from the aortic arch. Furthermore, the right vertebral artery did not originate from the right subclavian artery. Instead, at the level of the sternum manubrium, anteriorly to the trachea, the right vertebral artery branched off the right common carotid artery as its first branch. Subsequently, the right vertebral artery entered the transverse foramen at the level of C2. The left subclavian and common carotid arteries branched off normally from the aortic arch, and presented with no abnormalities in their anatomy. The diameters of the aortic arch and its branches were measured using a caliper. All measurements were taken two times by two independent researchers (WP and PO) and a mean value was established taking both measurements into account. The diameter of the ascending aorta at its origin was set at 30.4 mm. The diameter of the right subclavian artery at its origin was found to be 12.9 mm. The diameter of the right subclavian artery when it courses posteriorly to the trachea and esophagus was set at 9.9 mm. The diameter of the right subclavian artery where it gave off the costocervical trunk was found to be 8.4 mm. The aforementioned measurements, and additional diameters of the branches of the right subclavian artery are presented in Table 1.

Ethical concern

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The material used for this research was obtained from a body donation program.

DISCUSSION

The right subclavian artery usually develops from the right fourth aortic arch and seventh cervical intersegmental artery. However, the ARSA results from the regression of the right arch, between the right subclavian and right common carotid arteries, including the right ductus arteriosus. Subsequently, the proximal right subclavian artery arises from the distal right dorsal aorta, rather than from the right fourth aortic arch. This forms the retroesophageal portion of the ARSA [1, 8].

The course of the ARSA is quite variable. In most cases, it courses behind the esophagus and the trachea. However, the artery can also pass between the trachea and esophagus, and rarely, also anteriorly to the trachea [20]. The current case presents an ARSA with the most frequently seen retroesophageal course. Natsis et al. conducted a retrospective cadaveric study about the ARSA [20]. In the study, they reported that the bicarotid trunk with an ARSA is present in 0.16- 19.2% of the cases. However, in the present case, the carotid arteries originated independently from the aortic arch.

The ARSA is associated with other vascular and neural anatomic variations. A prime example is the Kommerells diverticulum, which is a remnant of the dorsal aortic arch [7]. Kommerell was the first to describe this anomaly in 1936, as a retroesophageal outpouching at the origin of an ARSA [13]. However, the aforementioned retroesophageal outpouching at the origin of an aberrant subclavian artery, is actually more prevalent in an aberrant left subclavian artery with a right aortic arch [2, 25]. Nevertheless, this vascular anomaly was not found in the present case study. The presence of an ARSA is also quite common in disorders such as Down's, and Edwards' syndromes, amongst others [28]. Furthermore, aberrant subclavian arteries are also associated with other cardiac anomalies, such as the tetralogy of Fallot [19]. Nakajima et al. reported that the incidence of an aberrant subclavian artery in patients with tetralogy of Fallot was 6% [19].

Variations of the branching pattern in aberrant subclavian arteries have been discussed in the literature. Keiffer et al. presented a study about the surgical treatment in patients with aberrant subclavian arteries [12]. In the study, they highlighted the coexistence of an ARSA with a vertebral artery originating from the right common carotid artery. Interestingly, this phenomenon was also observed in the present cadaver. Other abnormalities in the branching pattern of the subclavian artery were noted. The costocervical trunk had a more inferior origin in the thoracic cavity, more specifically, originating at the level of the 1st intercostal space, rather than posteroinferiorly to the clavicle. The branching pattern of this arterial trunk has been proven to be quite variable [4]. However, no abnormalities considering the branches of the costocervical trunk were observed in the present case. Furthermore, other studies have presented cases where the thyroid ima artery originated from the ARSA [12, 20].

An ARSA can compress the respiratory pathways, resulting in an increased risk of pulmonary infections and cyanosis [23]. However, respiratory symptoms in adults with an aberrant subclavian artery are rare. Polguj et al. stated that the reason for the different clinical manifestations of this anomaly is the difference in the anatomy of the trachea [23]. In an infant, the trachea is more compressible, and therefore, more prone to damage that can be caused by an ARSA. However, the adult trachea is more rigid and with that more resistant to compression by an aberrant subclavian artery. The sudden occurrence of symptoms in adults has been associated with age-related morphological alterations of the vessel due to atherosclerosis, and fibromuscular dysplasia among others [20]. Furthermore, in cases where stenosis or tortuosity of the aberrant subclavian artery is present, unequal upper extremity blood pressure, splinter hemorrhages, or vertebrobasilar ischemia might be present [21]. Patients with an ARSA that arises from the Kommerell diverticulum are also more susceptible to the development of an aneurysm [30]. The rupture of the said aneurysm is a life-threatening complication, and therefore, early detection is very important.

Gross performed the first successful surgical repair of an ARSA in 1946 on a fourmonth-old infant [6]. Previously, ARSA, and other aortic arch anomalies were diagnosed by endoscopy or upper gastrointestinal barium imaging. Nowadays, CT- angiography or MRangiography the most common method for diagnosing vascular anomalies, such as the ARSA [14]. Initial management, for mild symptoms, includes lifestyle changes, such as taking smaller sips and chewing well. Use of proton pump inhibitors for acid reduction has also been described [18]. If the conservative treatment does not work, surgical management is advised. The surgical treatment of a symptomatic aberrant subclavian artery consists of both endovascular and surgical treatments. A surgical approach to a symptomatic ARSA consists of dissection of the vessel and end-to-end anastomosis to the unilateral carotid artery [30]. However, Morris et al. presented a hybrid technique using both an endovascular approach with the use of an Amplatzer plug, and a surgical carotid subclavian bypass procedure to treat patients with an ARSA and dysphagia lusoria [18]. In the case study, the patient that underwent the said procedure recovered fully without any neurological deficits.

CONCLUSIONS

The present study presents a case of an ARSA, which is a rare anatomical variant of the branches of the aortic arch. The course and branching pattern of an aberrant subclavian artery is quite variable, and each variant can be associated with different possible complications. Furthermore, the ARSA is associated with other cardiovascular anomalies, such as the Kommerells diverticulum. Therefore, knowledge about the possible variations of this anomaly may be of great importance for physicians who encounter this variant in their practice.

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Conflict of interest: None declared

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Diameters	Mean Value (mm)
Ascending aorta at its origin	30.4
Right subclavian artery at its origin	12.9
Right subclavian artery when it courses posteriorly to the trachea and esophagus	9.9
Right subclavian artery where it gave off the costocervical trunk	8.4
Right costocervical trunk	2.1
Right thyrocervical trunk	4.4
Right common carotid artery at its origin	10.3
Right vertebral artery at its origin	3.8
Left common carotid artery at its origin	8.2
Left subclavian artery at its origin	8.3
Left vertebral artery at its origin	3.9
Left costocervical trunk	3.3
Left thyrocervical trunk	4.5

Table 1. Mean results of the measurements. Mm – millimeters.

Figure 1. Scheme presenting Aberrant Right Subclavian Artery (ARSA).

Figure 2. Subclavian artery and its close anatomical area – the Aberrant Right Subclavian Artery (ARSA) from the superior view.

Figure 3. A photograph of the Aberrant Right Subclavian Artery (ARSA) – an inferior view.



Thyroid gland Right lobe

Right subclavian a.

Omohyoid m.

Anterior scalene m.

Right vagus n. Right vertebral a.

Top

Right common carotid a.

Esophagus

Eches

Right brachiocephalic v.



