Chapter 5

The Epidemiological, Morphological, and Clinical Aspects of the Aberrant Right Subclavian Artery (Arteria Lusoria)

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Additional information is available at the end of the chapter

http://dx.doi.org/10.5772/64604

Abstract

The most common embryologic abnormality of the aortic arch is aberrant right subclavian artery (ARSA), known clinically as arteria lusoria (AL). This vessel travels to the right arm, crossing the middle line of the body and usually passing behind the esophagus. If the artery compresses the esophagus, it may produce a condition called dysphagia lusoria. Another commonly reported symptoms related to compression of adjacent structures by arteria lusoria were dyspnea, retrosternal pain, cough, and weight loss greater than 10 kg over a 6-month period. The chapter includes information describing demographic, clinical, and morphological characteristics of presence of arteria lusoria such as gender distribution, frequency in population, frequency of the most commonly reported symptoms related to compression of adjacent structures, coexistence with the most common vascular anomalies and diagnostic procedures. The presence of arteria lusoria together with the right nonrecurrent inferior laryngeal nerve (NRILN) is especially clinically important; during thyroid surgery, the right laryngeal nerve cannot be found at the lower pole of the thyroid, and it may be injured by the surgeon if it is not identified in the aberrant area or found lateral to the thyroid.

Keywords: aberrant right subclavian artery, arteria lusoria, anatomical variations, vessels, clinical symptoms

1. Introduction

The one of the most common embryologic vascular abnormalities of the aortic arch is an aberrant right subclavian artery (ARSA), known clinically as arteria lusoria (AL) [1].

In approximately 80% of individuals, three arteries arise from the arch of the aorta. From right to left, the brachiocephalic trunk arises (divided into the right common carotid artery and the right subclavian artery), followed by the left common carotid artery, and finally the left subclavian artery (**Figure 1**) [2].

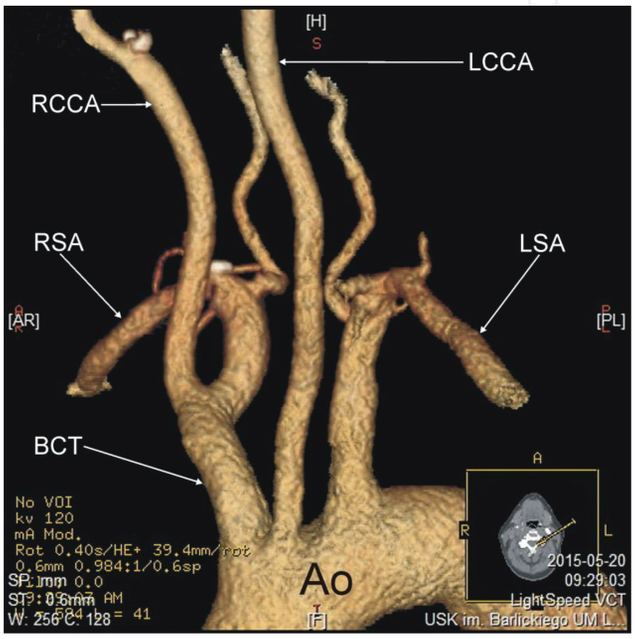

Figure 1. Three-dimensional computed tomography reconstruction of the arteries (CT-64-row MDCT scanner, Lightspeed VCT, GE, Waukesha, Wisconsin, USA). Ao—arch of the aorta, BCT—brachiocephalic trunk, LSA—left subclavian artery, LCCA—left common carotid artery, RCCA—right common carotid artery, RSA—right subclavian artery.

When an aberrant right subclavian artery (arteria lusoria) is present, the brachiocephalic trunk is absent and four arteries arise from the arch of the aorta: the right common carotid artery followed by the left common carotid artery, the left subclavian artery, and finally the right subclavian artery, with the most distal left-sided origin (**Figures 2–4**). This vessel, the aberrant right subclavian artery, travels to the right arm, crossing the middle line of the body and usually passing behind the esophagus (**Figure 3**) [3, 4]. If the artery compresses neighboring structures or organs, it may produce symptoms, the most common example being compression of the esophagus by the arteria lusoria, which results in a condition called dysphagia lusoria [4].

Although the first description of an aberrant right subclavian artery was provided in 1735 by Hunauld, the clinical entity was presented later [2]. In 1794, David Bayford, a physician from London, England, described a 33-year-old woman who succumbed to malnutrition after 20 years of progressive dysphagia [3]. At autopsy, Dr. Bayford noted esophageal compression by an abnormal right subclavian artery and suggested the term "dysphagia lusoria" to describe

http://dx.doi.org/10.5772/64604

this syndrome. Hence, it is also known as Bayford–Autenrieth dysphagia [3]. Kommerell described its radiological findings in 1936 [4].

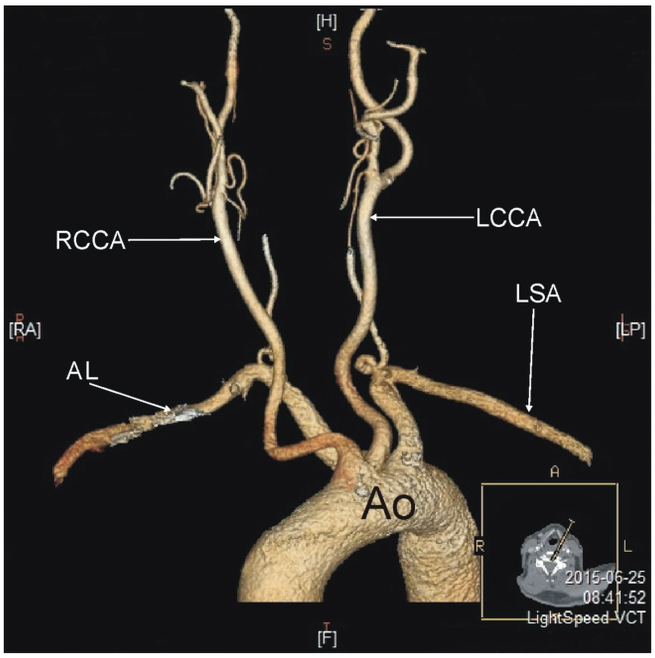

Figure 2. Three-dimensional computed tomography reconstruction of the arteries (CT-64-row MDCT scanner, Lightspeed VCT, GE, Waukesha, Wisconsin, USA). Ao—arch of the aorta, AL—arteria lusoria, LSA—left subclavian artery, LCCA—left common carotid artery, RCCA—right common carotid artery.

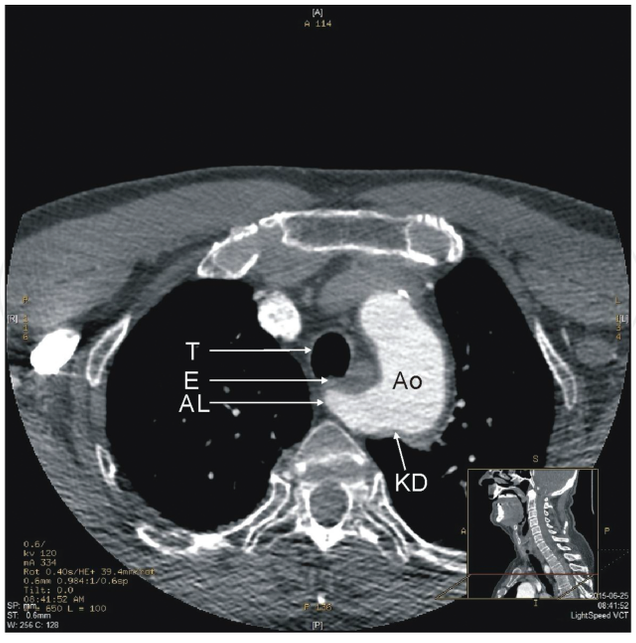

Figure 3. Computed tomography transverse scan on the level of arch of the aorta. Ao—arch of the aorta, AL—arteria lusoria, E—esophagus, KD—Kommerell's diverticulum, T—trachea.

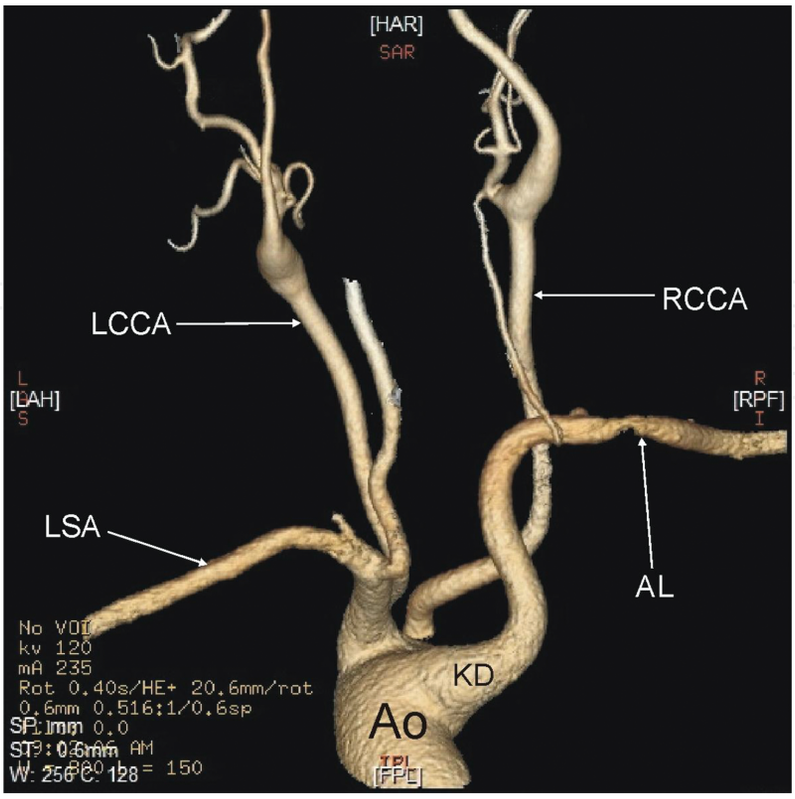

Figure 4. Three-dimensional computed tomography reconstruction of the arteries (CT-64-row MDCT scanner, LightSpeed VCT, GE, Waukesha, Wisconsin, USA). Ao—arch of the aorta, AL—arteria lusoria, KD—Kommerell's diverticulum, LSA—left subclavian artery, LCCA—left common carotid artery, RCCA—right common carotid artery.

2. Embryology

Arteria lusoria results from abnormal embryologic development of the aortic arch. In the normal situation, between the 4th and 5th weeks of embryonic life, blood leaves the heart by a common trunk called the "truncus arteriosus", which divides into two branches termed the ventral aortae. These branches are connected with the paired dorsal aortae by six aortic arches. The carotid system is formed by segments of the first three arches. The right fourth arch, a segment of the right ventral aorta, and a portion of the right dorsal aorta develop into the right subclavian artery. The left fourth arch persists as the adult aortic arch with the anlagen of the seventh dorsal intersegmental artery, and this forms the left subclavian artery. The fifth arches are both resorbed, and the sixth arches form the pulmonary artery and the ductus arteriosus [5, 6].

The aberrant origin of the right subclavian artery is caused by the involution of the right fourth vascular arch and proximal right dorsal aorta and the persistence of the seventh intersegmental artery originating from the proximal descending thoracic aorta, resulting in the arteria lusoria following an abnormal course [5, 6].

http://dx.doi.org/10.5772/64604

3. Epidemiological and demographic characteristics

The frequency of ARSA varies throughout the world. In Europe, depending on the country, it has been found in 0.11% (Great Britain) [7], 0.16% (Greece) [8], 0.3% (France) [9], or 0.36% (the Netherlands) [10] of the population. Studies have also been performed on other continents: Asia, 0.1–0.2% of cases (respectively: China and Japan) [11, 12]; North America, 0.5% of cases (United State) [13]; or Australia and Oceania, 0.8% of cases (New Zealand) [14] (**Table 1**). However, its detection arguably depends primarily on the sensitivity of the diagnostic procedures employed, for example, cadaveric or CT versus chest X-ray examination.

AL (%)	Country	Type of investigation	N/(total)	Researcher
0.1	China	Angiography	3/3000	Nie et al. [11]
0.11	Great Britain	Gastroscopy	1/920	Kelly [7]
0.16	Greece	Angiography	1/622	Natsis et al. [8]
0.2	Japan	Autopsy	1/516	Saito et al. [12]
0.3	France	coronarography	11/3730	Abhaichand et al. [9]
0.36	The Netherlands	Endoesophageal ultrasonography	12/3334	De Luca et al. [10]
0.5	USA	Computed tomography	36/7174	Haesemeyer and Gavant [13]
0.8	New Zealand	Autopsy	19/2291	Cainey [14]

AL—arteria lusoria.

Table 1. Frequency of aberrant right subclavian artery (arteria lusoria) in different populations.

The gender distribution of the aberrant right subclavian artery was found to be similar: 55.3% females versus 44.7% males [15].

4. Clinical characteristics

An aberrant right subclavian artery is usually asymptomatic, while about 8–10% of adult patients develop symptoms [16]. The anomaly does not cause symptoms in most patients and can be discovered incidentally during life or found at autopsy [17, 18].

When present, symptoms usually occur at the two extremes of life [19]. In children, respiratory symptoms are prevalent, mainly dyspnea or chronic coughing. They can also present with repetitive respiratory infections. Infant patients may demonstrate an increased frequency of pulmonary infections, which is thought to be due to absence of tracheal rigidity in combination with dysphagia and aspiration [19, 20]. As noted above, a rare cause of dysphagia observed in adults is compression of the esophagus by an abnormal course of the aberrant right subclavian artery, which is classically termed "dysphagia lusoria" [3].

Although dysphagia may be the most frequent symptom demonstrated by adults, children demonstrate a different respiratory symptomatology attributed to lack of the tracheal rigidity associated with dysphagia and false routes [21]. In addition, the arteria lusoria can also be revealed by the extension of aortic dissection or by peripheral arterial embolism.

A meta-analysis by Polguj et al. found the most commonly reported symptoms related to compression of adjacent structures by an aberrant right subclavian artery to be dysphagia (71.2%), dyspnea (18.7%), retrosternal pain (17.0%), cough (7.6%), and weight loss greater than 10 kg over a six-month period (5.9%) [15]. Among the less common symptoms, stomachache, back pain, and numbness of the right upper limb were reported. The mean age of the onset of symptoms was 49.9 ± 19.4 years for the whole group (data shown as mean ± standard deviation). However, the mean age according to gender was 44.9 ± 18.1 years for males and 54.0 ± 19.6 years for females. This difference was statistically significant [15].

Dysphagia also frequently occurs in elderly patients, for which four mechanisms have been proposed: increased rigidity of the trachea leading to easy compression of esophagus, aneurysm formation, presence of Kommerell's diverticulum, elongation of the aorta, the coexistence of an aberrant artery with a truncus bicaroticus, or a close origin of common carotid arteries from the arch of the aorta [22–25].

To the angiographer who uses the right axillary, brachial or radial approach to the ascending thoracic aorta, the arteria lusoria is also a clinically important element. The presence of an ARSA is suspected in cases in which catheterization of the ascending aorta proves difficult. Using the right radial approach, access to the ascending aorta is usually easy [11]. Previous studies indicate that only 60% of such cases were successfully performed by transradial approach in the setting of AL [26]. This variant makes the right transradial route difficult to approach the ascending aorta, as it requires the catheter to curve back to reach the aortic root [26, 27]. However, the repeated entry of the guide wire from the right subclavian artery to the descending aorta rather than the ascending aorta should indicate this possibility. Thus, angiography can prove to be very challenging in the presence of an arteria lusoria [11, 26, 27].

Finally, the inferior right recurrent laryngeal nerve is an asymptomatic variation anomaly, which can be an important obstacle and be seriously damaged during cervicotomy, thyroid, and parathyroid surgery. In such cases, the inferior right recurrent laryngeal nerve is a classic risk and must be eliminated by location and routine dissection of the nerve [28–30]. This is of particular importance when the diagnosis concerns an asymptomatic neural anomaly discovered by dissection or a vascular anomaly whose symptoms are very variable [30].

5. Morphological characteristics

The literature presents two main classifications of the aberrant right subclavian artery. According to Neuhauser's threefold classification, the first type of arteria lusoria crosses the posterior wall of the esophagus, and this is observed in more than 80% of the cases. In the

http://dx.doi.org/10.5772/64604

second type, this artery passes between the trachea and the esophagus (15% of the cases), and in the third type, it crosses the midline of the body ahead of the trachea (in 5% of the cases) [23].

In contrast, the Adachi and Williams classification recognizes four basic morphological types. Type I/G is characterized by an aberrant right subclavian artery arising from the arch of the aorta as the final branch. Type II/CG is similar to the first type, but an additional left vertebral artery arises from the arch of the aorta. In type III/H, three arteries arise from the arch of the aorta: as the first common trunk of the common carotid arteries (truncus bicaroticus), as the left subclavian artery, and as the last aberrant right subclavian artery. In type IV/N, the aberrant left subclavian artery arises from the right-sided arch of the aorta as the final branch [31].

The most common vascular anomalies coexisting with an aberrant right subclavian artery (arteria lusoria) were found to be truncus bicaroticus (19–29%), Kommerell's diverticulum (15–60%), aneurysm (just after the origin of arteria lusoria) (13%), and right-sided aortic arch (9%) [15]. Klinkhamer regards truncus bicaroticus is a precondition for tracheal–esophageal compression and the development of clinical symptoms. Under these circumstances, the truncus bicaroticus holds the trachea from the front, and the aberrant right subclavian artery compresses the esophagus from behind [22].

In 1936, Kommerell published the first radiological findings of the route of the aortic arch as an aortic diverticulum (Kommerell's diverticulum), which was identified as being located at the origin of an aberrant subclavian artery [4] (**Figure 3** and **4**). Kommerell's diverticulum is usually found incidentally on a chest roentgenogram and is often misdiagnosed as a mediastinal tumor [29]. Kommerell's diverticulum is a normal broadening of the proximal origin of the aberrant right subclavian artery from the aortic arch and is most frequently present in patients with a right aortic arch and an aberrant left subclavian artery [32]. However, Kommerell's diverticulum is not analogous with an aneurysm: The primary indication for surgical repair of Kommerell's diverticulum is a diameter larger than 50 mm and the presence of clinical symptoms.

The identification of arteria lusoria should alert the radiologist and surgeon that a nonrecurrent inferior laryngeal nerve (NRILN) is present and that an anticipating surgical technique should be performed to reduce the risk of neural injury. Due to its anatomical position, an NRILN is not only at risk of being damaged during thyroidectomy, but also during such other surgical procedures as neck dissection, parathyroidectomy, and carotid endarterectomy [30, 33].

6. Diagnosis

The diagnosis of arteria lusoria was reported only on anatomical dissection until 1936, when Burckhard Kommerell described the clinical diagnosis of an aberrant right subclavian artery that originated from an aortic diverticulum, later known as Kommerell's diverticulum, in a 65-year-old man who was believed to have stomach cancer [4]. In 1946, Gross was the first to report the surgical treatment of dysphagia lusoria, in a four-month-old infant [34].

The diagnostic modalities available to visualize an arteria lusoria include barium esophagogram, computed tomography (CT), magnetic resonance imaging (MRI), digital subtraction angiography (DSA), endoscopy and endoscopic ultrasound. New advances in CT technology allow even small vascular structures to be visualized in detail. Multidetector computed tomography (MDCTA) is now an established diagnostic test in the evaluation of many vascular diseases [35–39].

Barium contrast examination of the esophagus shows a characteristic, extrinsic, smooth diagonal impression at the level of the third and fourth dorsal vertebra. Lateral or oblique views show the extrinsic impression to be posterior, and in case of arteria lusoria, just above the level of the aortic arch. As dysphagia occurs frequently with ingestion of solid foods, including a barium soaked bread bolus may improve localization of the defect [35].

Digital subtraction angiography gives valuable information regarding AL. It is an invasive procedure and, in contrast to MDCT, has the disadvantage in showing extravascular structures such as the esophagus. It has also been shown that the effective radiation doses in MDCT angiography studies are moderate and even lower than those associated with DSA in a comparable patient group.

CT or MRI (magnetic resonance imaging) angiography has replaced conventional angiography and is the gold standard for the diagnosis. It not only confirms the diagnosis but also helps to exclude aneurysm of the aorta or other associated anomalies and to plan the operation [36–39].

MRI has the advantage of being a noninvasive procedure and the patient is spared the potential risk of intravenous contrast agents. MRI is not as useful as MDCT due to the generation of respiratory and cardiac motion artifacts. Also it is not a preferred method due to its cost and prolonged scan time. Although MR angiography may reveal the presence of a vascular anomaly, the information regarding nonvascular mediastinal structures is insufficient [36–39].

Endoscopy may reveal pulsatile, shelf-like extrinsic compression in the posterior wall of the esophagus, with intact mucosa. Such an area of narrowing is usually located between 20 and 24 cm from the mouth [7].

Endoscopic ultrasound (EUS) can identify an arteria lusoria, as it lies close to the esophagus. EUS is regarded as the most accurate test for the evaluation of the esophageal wall and the surrounding structures, with the incorporation of Doppler technology in modern echoendoscopes allowing particularly accurate examination of adjacent vessels [10].

The combination of MDCT with 3D volume rendering images provides further advantages. These allow not only the depiction of the thoracic vascular anomalies but also more accurate assessment of the diameter, angle, and compressed area of the esophagus and the relationship between the AL with the esophagus and other mediastinal structures. In addition, MDCT is a noninvasive procedure, unlike DSA, and offers easier application and a shorter time requirement than DSA or MRA [36–39].

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7. Conclusion

A familiarity with the anatomy of the some types of vascular anomalies is necessary for clinicians involved in many medical areas. Compression of adjacent structures by an arteria lusoria needs to be differentiated from other conditions presenting symptoms such as dysphagia, dyspnea, retrosternal pain, cough, and weight loss. The knowledge presented in this chapter will allow the best healthcare to be provided for patients.

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