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A bicarotid trunk in association with an aberrant right subclavian artery. Report of two cases, clinical impact, and review of the literature

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A combination of an aberrant right subclavian artery (ARSA) and a bicarotid trunk (BCT) appears in up to 2.5% of the population. The aim of this study is to report the higher total and male incidence of this variation in the literature and to summarise its clinical impact, providing useful knowledge to anatomists, radiologists, cardiologists, and vascular and thoracic surgeons in order to avoid diagnostic pitfalls and therapeutic complications. A total of 72 (43 female and 29 male) Greek Caucasian formalin-embalmed cadavers were studied. The international literature was reviewed along with the dissection archives of the Department of Anatomy from 1986 to 2009. Two male cadavers were found to have an ARSA combined with a BCT (incidence: total 2.78%, males 6.9%, females 0%). Both aortic arches consisted of three branches: (1) the BCT, (2) the left subclavian artery, and (3) the ARSA. The common carotids followed a normal route to the neck; the ARSA passed between the trachea and the oesophagus in the first case and behind the oesophagus in the second case, and was accompanied by a non-recurrent laryngeal nerve. In the second cadaver the ARSA formed a sharp angle (kinking/buckling) on its route to the right arm. The ARSA is associated with several congenital cardiovascular anomalies and some chromosomal and other syndromes. It is occasionaly responsible for causing dysphagia, dyspnoea, or acute ischaemia to the right upper limb, and it may present as a superior mediastinal mass in cases of aneurysm formation. (Folia Morphol 2011; 70, 2: 68–73)

Key words: aortic arch variations, common origin of carotids, dysphagia

INTRODUCTION

The right subclavian artery normally arises from the brachiocephalic trunk and is the first branch to come off the ascending aorta. An aberrant right subclavian artery (ARSA) arises as the last branch of the left-sided aortic arch, distal to the normally positioned left subclavian artery [15]. Asherson [2] cited Bayford who described a patient with dysphagia secondary to oesophageal compression by an ARSA and coined the term "dysphagia lusoria". In its course to the right arm it usually (80%) crosses the midline behind the oesophagus and less frequently between the trachea and oesophagus (15%) or in front of the trachea (5%) [5]. Its relative frequency varies from 0.1% to 4% accord-

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ing to different authors and among different study materials (autopsy, dissection, and radiologic findings) [3, 13, 24]. It is associated with several congenital cardiovascular anomalies and some chromosomal and other syndromes [29] as well as with the presence of a right non-recurrent inferior laryngeal nerve [4]. The bicarotid trunk (BCT), another aortic arch branching variation, is a trunk that arises from the aortic arch and then bifurcates into the right and left common carotid arteries, thus creating a common origin for the common carotid arteries. A combination of an ARSA and a BCT appears in up to 2.5% of the population [14, 22, 24, 28]. ARSA usually remains asymptomatic and is revealed incidentally either radiologically or during anatomy dissection. However, it is rarely responsible for causing dysphagia, dyspnoea, or acute ischaemia to the right upper limb, and it may present as a superior mediastinal mass in cases of aneurysm formation [7, 17]. The aim of this study is to describe two cases of ARSA in association with BCT, among 72 dissected cadavers, and to refer to its incidence and clinical impact according to literature reports. This information will provide useful knowledge to anatomists, radiologists, cardiologists, and vascular and thoracic surgeons.

MATERIAL AND METHODS

During routine dissection for educational purposes in the Department of Anatomy of the Medical School of the Aristotle University of Thessaloniki, from 1986 to 2009, a total of 72 (43 female and 29 male) Greek Caucasian formalin-embalmed cadavers were studied. The literature was reviewed along with the dissection archives of our department from 1986 to 2009.

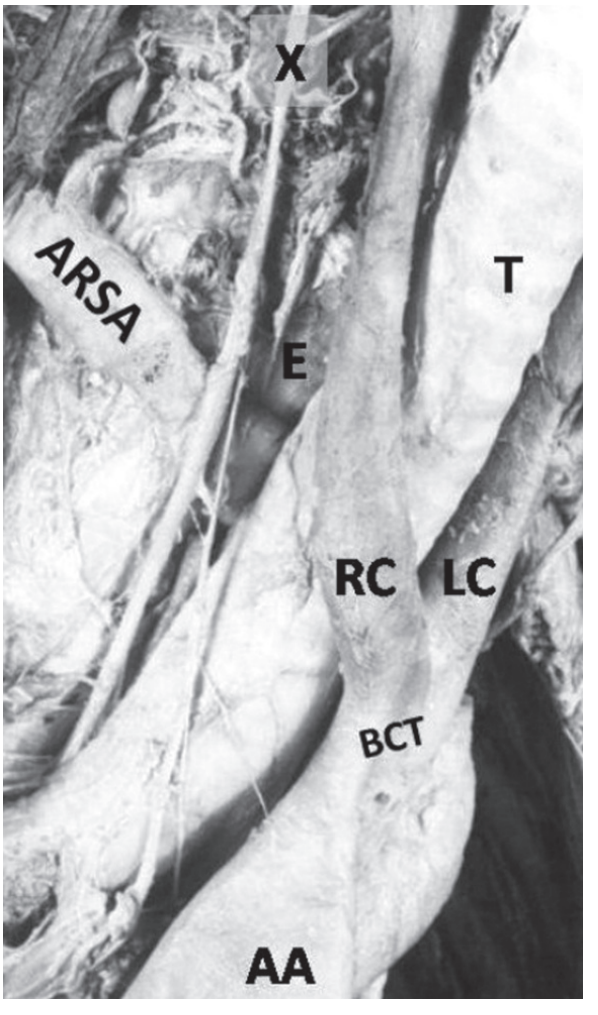

Figure 1. Case 1. Right view of the aortic arch and its branches showing the bicarotid trunk (BCT) and the route of the aberrant right subclavian artery (ARSA) between trachea and oesophagus; AA — aortic arch; RC — right common carotid artery; LC — left common carotid artery; T — trachea; E — oesophagus; X — vagus nerve.

RESULTS

Two cadavers were found to have an aberrant right subclavian artery combined with a BCT and are described in detail below.

Case 1 (Fig. 1); Caucasian, male, 76 years old, cause of death: pulmonary tuberculosis.

An aortic arch with three branches was observed. The first branch from the aortic arch was not the brachiocephalic trunk (innominate artery) but a trunk (height: 34 mm, diameter: 25 mm) that divided into the right and the left common carotid arteries (BCT). The common carotids followed a normal route to the neck after their common origin. The second branch was the left subclavian artery also following a normal route to the left upper limb. The last branch was the aberrant right subclavian artery (diameter: 12 mm, level of origin: 4th thoracic vertebra). It followed a slightly rightward and upward route and coursed between the trachea and the oesophagus. Its route to the right upper limb was normal. An unusual formation of the right vagus nerve was also observed with a non-recurrent right laryngeal nerve present. No aneurysmal dilatation of the ARSA or the aorta itself was observed, and the right vertebral artery maintained its subclavian origin with a normal distribution pattern. The thoracic duct followed a normal route to the neck but arched posterior to the ARSA and ended in the junction of the left subclavian and internal jugular veins. All other anatomical structures in the neck and thorax were according to the normal anatomical pattern, and

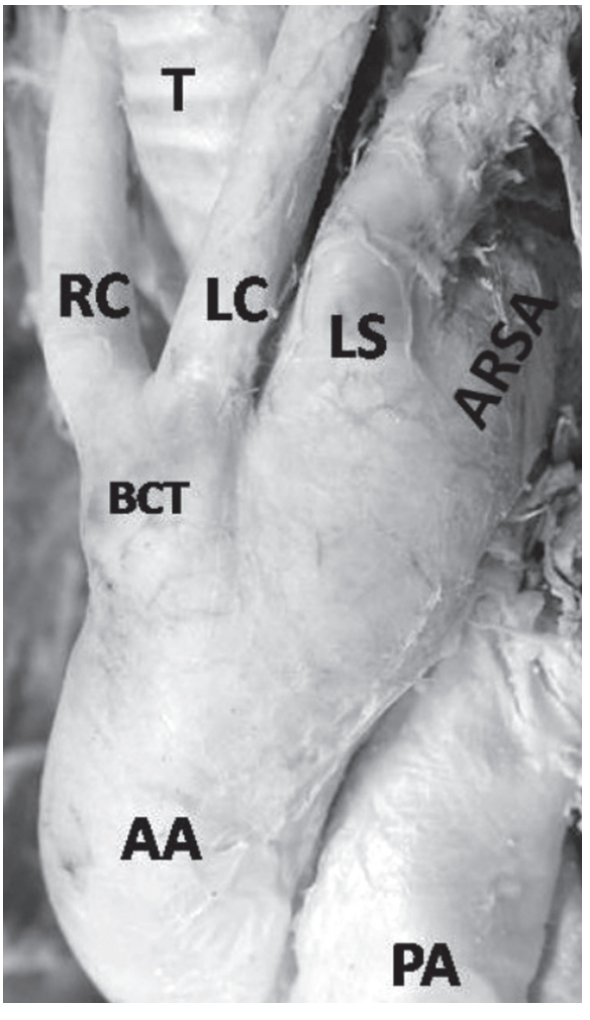

Figure 2. Case 2. Left view of the aortic arch and its branches showing the bicarotid trunk (BCT) and the origin of the aberrant right subclavian artery (ARSA); AA — aortic arch; RC — right common carotid artery; LC — left common carotid artery; T — trachea; LS — left subclavian artery; PA — pulmonary artery.

no relevant clinical symptoms were reported on the cadaver's medical history.

Case 2 (Fig. 2); Caucasian, male, 81 years old, cause of death: lung cancer (pancoast tumour of the left upper lobe).

An aortic arch with three branches was observed. The first branch from the aortic arch was the BCT (height: 30 mm, diameter: 24 mm) giving rise to the two common carotid arteries that followed a normal route to the neck after their common origin. As a second branch, close to the previous trunk, arose the left subclavian artery also following a normal route to the left upper limb. The last branch was the aberrant right subclavian artery (diameter: 16 mm, level of origin: 3rd thoracic vertebra) originating from the dorsal aspect of the aortic arch near the left subclavian artery. It followed a slightly rightward and upward route to the left edge of the oesophagus. There it coursed horizontally behind the oesophagus at the level of the 7th cervical vertebra. After crossing the oesophagus the ARSA turned upwards to the neck until it reached the upper edge of the thyroid gland (level of the 5th cervical vertebra). There it turned abruptly to the right forming a sharp angle (kinking/buckling) and was finally directed to the right arm. The angle was acute (80 degrees) with the right upper limb in adduction and obtuse (100 degrees) in abduction. It was accompanied by a right non-recurrent laryngeal nerve. No aneurysmal dilatation of the ARSA or of the aorta itself was observed and the right vertebral artery maintained its ordinary origin and pattern of distribution. The thoracic duct followed a normal route to the neck, looped anterior to the left subclavian (and the underlying ARSA) artery, and ended at the jugular-subclavian junction. No other vascular variations or associated cardiac anomalies were found. No relevant clinical symptoms were reported on the cadaver's medical history.

DISCUSSION

Variations in the branching of the human aortic arch are numerous. The usual/normal aortic arch gives rise to three branches: (1) the innominate artery (branching subsequently to the right subclavian and to the right common carotid artery), (2) the left common carotid artery, and (3) the left subclavian artery [15].

Anatomical variations associated with ARSA include the abnormal origin of the right vertebral artery from the aorta or the right common carotid artery, the presence of a common carotid trunk, a right-sided thoracic duct, and a right non-recurrent inferior laryngeal nerve [16].

Aberration from the normal embryologic development of the primitive aortas and aortic arches results in the formation of an ARSA [5]. The 4th vascular arch involutes with the right dorsal aorta, while the 7th intersegmental artery remains attached to the descending aorta. This persistent intersegmental artery becomes the right subclavian artery and leads to an aberrant artery, which often follows a retro-oesophageal course. An aortic diverticulum (Kommerell's diverticulum) may be present at the origin of this vessel representing the remnant of the distal right aortic arch. The ARSA usually arises from the dorsal margin of the aortic arch and in few cases from the descending aorta. The proximal part of the artery is generally greater in diameter than the distal part [13].

In addition, abnormal development of the 4th right aortic arch results in the right inferior laryngeal nerve stemming directly from the cervical part of the vagus nerve, without having a recurrent course to reach the cricothyroid membrane (non-recurrent inferior laryngeal nerve) [4]. There are two main types described: Type 1, the nerve arises directly from the cervical vagus and runs together with the vessels of the superior thyroid peduncle; and Type 2, it follows a transverse path parallel to the inferior thyroid artery (further subdivided into type 2A when it runs over the trunk and type 2B when it runs under the trunk or between the branches of the artery). This absence of the right recurrent laryngeal nerve, which has a frequency of 0.3––1.6%, is clinically important because if unrecognised, it may be susceptible to injury during surgery [4]. A potential compression of the thyroid gland by small tumours has also been reported [15]. In our cases a Type 2B right non-recurrent laryngeal nerve was present following a transverse path parallel to and under the trunk of the right inferior thyroid artery.

The ARSA appears in the general population with an incidence of 0.1–4.0% [1, 13, 24]. The incidence of ARSA combined with BCT varies from 0 to 2.5%, being higher in the African race (Table 1) [14, 22, 28]. In Caucasian specimens, Williams et al. [28] found it once in 80 Caucasian cadavers, and it was not found by DeGaris et al. [14] and McDonald and Anson [22] in over 100 Caucasian cadavers. We found two cases in 72 Greek Caucasian cadavers, thus having an incidence of 2.78%, higher than that found by any other author, even among African people. There are only two reports in the literature describing this variation twice in their specimens, and there is no study describing more than two cases [14, 28]. This incidence is also much higher compared with our former study [24] on angiographies (0.16%: 1 case in 633 angiographies), reflecting the difference in the study material. Most studies revealed a higher incidence of the variation in females than in males [14, 22, 28] while both our studies [24, present study] show higher rates in males. This combined anomaly has also been presented in some case reports found either incidentally or after investigation for certain symptoms [3, 7, 10, 16, 17, 26].

The prevalence of ARSA is greater in patients with Down syndrome (ranging from 3.6% to 37%) and with chromosome 22q11 deletion (CATCH 22), trisomy 18, Noonan syndrome, and Potter syndrome. The incidence in patients with congenital heart and arterial anomalies such as Fallot's tetralogy or pulmonary atresia is also higher than in the general population [11, 29].

The ARSA is usually asymptomatic, as it was in our cases according to the cadavers' medical histories. However, rarely, several symptoms and clinical manifestations have been attributed to it, like dysphagia, dyspnoea, recurrent pulmonary infections, feeding difficulty, aneurysm formation, and acute ischaemia of the right arm. The term "dysphagia lusoria" or difficulty in swallowing due to a trick of nature (aberrant artery) was introduced by Bayford in 1794 [2]. This situation is characterised by difficulty in swallowing and pain in some cases, due to the pressure applied by the artery onto the wall of the oesophagus [17]. In this case, surgery is necessary [23]. Furthermore, it may cause serious respiratory symptoms, without the presence of dysphagia in paediatric patients. In children, tracheoesophageal compression manifests itself by respiratory symptoms, while in adults the trachea is more rigid and more resistant to compression, leaving dysphagia as the principal symptom [17]. When present in front of the trachea it may cause complications during tracheostomy [9]. It is supported that compression occurs: (1) if the common or close origin of the carotid arteries is such that anterior evasion of the trachea is prevented, (2) if the space around the trachea and oesophagus is narrow enough to prevent an escape from compression in any direction, and (3) when advanced atherosclerosis results in rigid vessels that can compress the oesophagus [20].

Aneurysm and dissection of an ARSA have been reported with an incidence of 3–8% [8, 12]. They appear as a superior mediastinal mass, either asymptomatic or causing dyspnoea, cough, swallowing difficulty, regurgitation, hoarseness, torticollis, vertigo, retrosternal pressure, and pain referring to the right arm [19]. In addition, the walls of an ARSA are thinner than those of normal arteries. All this poses a high risk of spontaneous rupture or perforations of the oesophagus into an ARSA by foreign body or rupture of the aneurysm into the oesophagus. Additional risks of thromboembolism, rupture into the mediastinum or pleura, and even superior vena cava syndrome exist [18, 19]. About half of the cases of aneurysmal ARSA reported in the literature ruptured with conservative management, prompting some authors to recommend surgery or stenting even in asymptomatic patients [12].

When stenosis or kinking/buckling of the aberrant vessel is present, the findings may include unequal upper extremity blood pressure readings, right arm claudication, splinter haemorrhages, or vertebrobasilar ischaemia. In this case, physical examination and

Table 1. Studies and case reports pertaining to an aberrant right subclavian artery (ARSA) in combination with bicarotid trunk (BCT)

Study	Incidence	Race/material	Sample size	Total cases of ARSA+BCT	Symptoms
Williams et al. (1932)	Total: 2.5% In males: 1.8% In females: 4.3%	African/cadavers	Total: 79 Males: 56 Females: 23	Total: 2 Males: 1 Females: 1	Not mentioned
	Total: 1.3% In males: 0% In females: 12.5%	Caucasian/cadavers	Total: 80 Males: 72 Females: 8	Total: 1 Males: 0 Females: 1	Not mentioned
DeGaris et al. (1933)	Total: 1% In males: 0% In females: 3.1%	African/cadavers	Total: 203 Males: 138 Females: 65	Total: 2 Males: 0 Females: 2	Not mentioned
	Total: 0% In males: 0% In females: 0%	Caucasian/cadavers	Total: 111 Males: 98 Females: 13	Total: 0 Males: 0 Females: 0	Not mentioned
McDonald & Anson (1940)	Total: 1.7% In males: 0% In females: 12.5%	American-African/ /cadavers	Total: 59 Males: 51 Females: 8	Total: 1 Males: 0 Females: 1	Not mentioned
	Total: 0% In males: 0% In females: 0%	American-Caucasian/ /cadavers	Total: 157 Males: 149 Females: 8	Total: 0 Males: 0 Females: 0	Not mentioned
Boas et al. (2002)	Case report	Not mentioned/ /angiography	Case report	79 year-old female	Acute ischemia of the right upper extremity
Epstein & DeBord (2002)	Case report	Caucasian/ /angiography	Case report	33 year-old male	Progressive dysphagia
Fazan et al. (2003)	Case report	Caucasian/ /cadaver	Case report	54 year-old female	Not mentioned
Poultsides et al. (2004)	Case report	Caucasian/ /cadaver	Case report	77 year-old male	Asymptomatic
Attmann et al. (2005)	Case report	Not mentioned/ /contrast enhanced computed tomography scan + angiography	Case report	75 year-old male	Asymptomatic aneurysm
Chahwan et al. (2006)	Case report	Not mentioned/ /angiography	Case report	75 year-old male	Transient ischemic attack
Natsis et al. (2009)	Total: 0.16% In males: 0.22% In females: 0%	Greek-Caucasian/ /angiographies	Total: 633 Males: 447 Females: 186	Total: 1 Males: 1 Females: 0	Not mentioned
Present study (1986–2009)	Total: 2.78% In males: 6.9% In females: 0%	Greek-Caucasian/ /cadavers	Total: 72 Males: 29 Females: 43	Males: 2 Total: 2 Females: 0	Asymptomatic

angiography may be useful to differentiate this condition from arterial thoracic outlet syndrome [21, 27]. Transient compression of an ARSA during transesophageal echocardiography imaging, while not life threatening, can impair haemodynamic monitoring if radial arterial catheters are used on the affected side [6]. An unsuspected ARSA is also at risk of accidental injury during any form of invasive procedure, such as thoracoscopic oesophagectomy [25].

Diagnosis of an ARSA can be made by plain chest X-ray, barium oesophagogram, angiography, computed tomography images, magnetic resonance images,

or angiographies but it is often an incidental finding during patient investigation for another reason [8].

It is clearly stated that, except from the pure anatomical interest, ARSA implies a significant clinical impact and should be borne in mind by general practitioners, radiologists, thoracic and vascular surgeons, interventional practitioners, and even cardiologists and gastroenterologists in order to avoid diagnostic pitfalls and therapeutic complications.

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