

Aberrant Right Subclavian Artery: A Cadaveric Case Study with Anatomy Suggestive of Subclavian Steal Syndrome

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ABSTRACT

Aberrant right subclavian artery (ARSA) is a congenital variation in which the right subclavian artery arises from the aortic arch distal to the left subclavian artery instead of branching from the brachiocephalic trunk. This anomaly has the potential to deviate the normal course of the right recurrent laryngeal nerve (RRLN) during embryological development to cause a non-recurrent laryngeal nerve (NRLN). This case report presents the finding of an ARSA in a 94-year-old female cadaver during routine dissection for medical education. A proximal stenosis of the right subclavian artery was discovered, indicating the possibility of subclavian steal syndrome (SSS), and a NRLN was present. Although often asymptomatic, SSS reduces blood flow to the brain and can cause many neurological symptoms. If left unidentified, a NRLN can have significant surgical complications, and is particularly pertinent to thyroidectomy and cervical spine procedures. This case report emphasizes the importance of awareness of this rare neurovascular anatomical variation, knowledge of the associated embryology, and screening for neurovascular anomalies in preoperative patients.

Keywords: Anatomy; Anomaly; Cadaver; Subclavian; Vasculature

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INTRODUCTION

Typically, the aortic arch gives rise to the brachiocephalic trunk, left common carotid artery, and the left subclavian artery. The brachiocephalic trunk divides into the right common carotid artery and the right subclavian artery. The right subclavian artery branches into the right vertebral artery, internal thoracic artery, and thyrocervical trunk. These arteries come from the left subclavian artery on the left side. The branches of the subclavian arteries supply blood to the upper limbs, neck, and posterior portion of the brain. The vertebral arteries enter the skull through the foramen magnum and unite to form the basilar artery. The basilar artery then joins the

internal carotid arteries through the posterior communicating arteries at the Circle of Willis [1].

The left recurrent laryngeal nerve and right recurrent laryngeal (RRLN) nerve branch from their corresponding vagus nerves. Typically, the RRLN loops inferior to the right subclavian artery and then ascends posterior to the artery toward the larynx in the tracheoesophageal groove. The left recurrent laryngeal nerve loops inferior to the aortic arch, passing posterior to the ligamentum arteriosum before ascending toward the larynx [2].

Common aortic arch variants include a bovine arch, a right-sided aortic arch, a left vertebral artery originating from the aortic arch, and an aberrant right subclavian artery. The bovine arch variant is a common variant and occurs when the left common carotid artery arises from the brachiocephalic trunk instead of directly from the aortic arch. In a right-sided aortic arch variant, the aortic arch curves to the right of the trachea and esophagus and passes over the right main bronchus, instead of the typical left-sided course. In the left vertebral artery variant, the left vertebral artery originates from the aortic arch between the left common carotid and left subclavian arteries, rather than branching from the left subclavian artery. An aberrant right subclavian artery (ARSA) is the most common congenital anomaly of the aortic arch. In this case, the right subclavian artery arises from the aortic arch distal to the left subclavian artery and passes posterior to the esophagus, instead of arising from the brachiocephalic trunk, making it the retroesophageal variant [3]. Other variants of aberrant subclavian arteries include a course in front of the trachea, and between the trachea and esophagus [4].

A right non-recurrent laryngeal nerve (NRLN) variant occurs when there is an ARSA. This causes the NRLN to lie superior to the aberrant artery and not loop around the vessel, instead traveling horizontally from the vagus nerve to the larynx [5].

In subclavian steal syndrome (SSS), an occlusion or stenosis of the subclavian artery results in retrograde blood flow through the ipsilateral vertebral artery to supply the arm. In essence, blood is redirected from the posterior cerebral circulation to the arm. Symptoms of SSS vary but may include arm claudication, pain, numbness, weakness, and coldness of the affected arm, as well as headache, dizziness, or syncope [6].

With the individual presented in this case report, the right subclavian artery is branching directly off the aortic arch with an associated NRLN. Thus, the aortic arch has four branches: the right subclavian artery, brachiocephalic trunk, left common carotid artery, and left subclavian artery. Additionally, there is a constriction of the right subclavian artery before the branching of the right vertebral artery, which is consistent with SSS.

CASE REPORT

An ARSA was identified upon dissection of the

upper thoracic cavity on a 94-year-old female cadaver at Liberty University College of Osteopathic Medicine (LUCOM). The ARSA was first observed after removing the anterior thoracic wall and reflecting the infrahyoid muscles superiorly. This was followed by the removal of the thymus via blunt dissection to expose the superior mediastinum. Grant's Dissector [7] was used to direct the dissection.

Once the superior mediastinum was exposed, the right common carotid artery and the ARSA could be seen arising from the aortic arch, and a brachiocephalic trunk was not present. The ARSA traveled posterior to the trachea and the esophagus. This position of the ARSA is considered a retroesophageal variant [4]. Additionally, a coarctation was identified in the ARSA just before it crossed the first rib, approximately 69.76mm distal to the aortic arch, and 11.5mm proximal to the branching of the vertebral artery. This stenotic narrowing appeared to restrict blood flow to the rest of the artery and caused dilation of the vessel, approximately 20.1mm in width, at the junction of the subclavian artery and the aortic arch. These measurements were obtained via caliper measurement after initial dissection, utilizing the Mitutoyo No. 505-646 caliper. Along with the ARSA presentation, the RRLN was presented as a NRLN variant. The NRLN branched off the vagus nerve and ran directly upward to enter the tracheoesophageal groove in the neck. The left recurrent laryngeal nerve and all other vasculature presented normally (Figures 1-3).

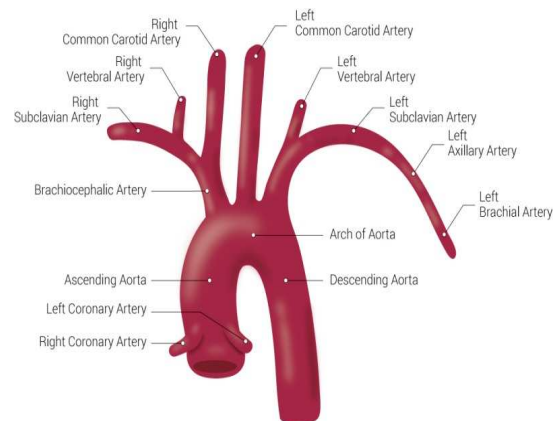


Figure 1: Typical aortic arch [8].

A typical aortic arch contains three direct branches. From left to right: brachiocephalic artery, left common carotid artery, left subclavian artery. The brachiocephalic artery gives rise to the right subclavian artery and right common carotid artery.

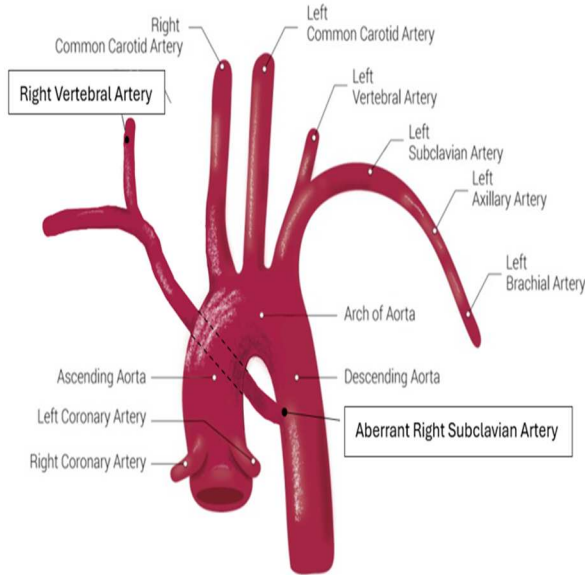


Figure 2: Illustration of Aberrant Right Subclavian Artery, adapted from [8].

This image attempts to illustrate the ARSA in comparison to the previous image by Shahoud, Miao, and Bolla [8]. Note the origin of the ARSA on the first part of the descending thoracic aorta. It courses posterior to the ascending aorta, as indicated by the dashed lines, and then continues superiorly.

A variation in the aortic arch was identified in a cadaver at LUCOM. The aortic arch (cut) is separated from the heart (removed) and oriented to place the right common carotid artery at the most superior position. The order of arteries from superior to inferior is the following: right common carotid artery, left common carotid artery, left subclavian artery, right subclavian artery. The right subclavian artery and the right common carotid artery are not connected via the brachiocephalic artery, as shown in Figure 1, as the right subclavian artery is branching from the distal portion of the aortic arch to the right side of the body.

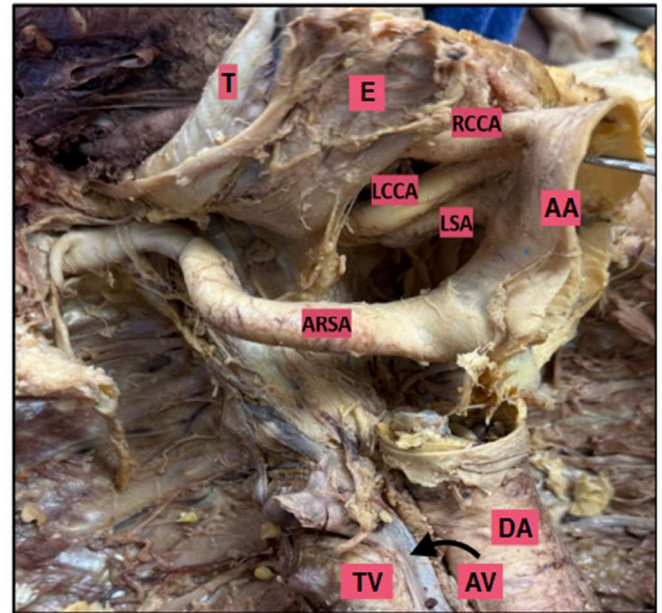


Figure 3: Aberration in right subclavian artery.

Legend: T: Trachea, E: Esophagus, RCCA: Right Common Carotid Artery, LCCA: Left Common Carotid Artery, LSA: Left Subclavian Artery, AA: Aortic Arch, ARSA: Aberrant Right Subclavian Artery, DA: Descending Thoracic Aorta, TV: Thoracic Vertebrae, AV: Azygos Vein

DISCUSSION

Embryology: Abnormalities in embryonic development explain both the ARSA, which is found in about 0.5-4.4% of the population [9], and the NRLN in this individual. After the fusion of the endocardial heart tube, the dorsal and ventral aortae are connected by pharyngeal arch arteries. In the third week of gestation, the aorta develops, receiving contribution from the left 4th arch artery [10], and part of the aortic sac [11]. In typical development, the proximal part of the right subclavian artery is derived from the right 4th pharyngeal arch artery, and the distal part is formed from the 7th cervical intersegmental artery. The left subclavian artery is derived fully from the 7th cervical intersegmental artery and the common carotid arteries are derived from the 3rd arch arteries [10]. In this way, the common carotid and subclavian arteries become connected to the aortic arch by the brachiocephalic trunk on the right, while these arteries branch directly from the aortic arch on the left side [10].

In this individual, however, the right common carotid artery and right subclavian branch directly from the aortic arch. This was likely due to abnormal regression of the right 4th pharyngeal arch artery. This could cause the right subclavian artery to be formed from only the distal portion of the right dorsal aorta and the right seventh cervical intersegmental artery, resulting in the right subclavian artery originating directly from the aortic arch [12]. The regression of the right 4th pharyngeal arch artery would also explain the NRLN present in this individual as abnormalities of the 4th arch artery would prevent the typical looping [13]. Thus, the NRLN nerve is associated with right subclavian artery abnormalities in most cases, as the proper development of the right subclavian artery is crucial to the appropriate course and formation of the RRLN [13].

Clinical Implications: The term “Subclavian Steal Syndrome” was first coined by C.M. Fisher in the New England Journal of Medicine after case reports by Contorni and Rievich in the early 1960s. While the exact prevalence of SSS is not known, studies have shown a 2.5% incidence rate of the population fulfilling the criteria [14]. Additionally, the syndrome remains characteristically asymptomatic, making the exact prevalence unknown and difficult to study [6].

The main cause of SSS is an occlusion of the subclavian artery proximal to the origin of the vertebral artery [15]. This syndrome can have many etiologies, with the main being atherosclerosis [16]. Other causes are attributed to Takayasu arteritis, subclavian artery compression in the thoracic outlet, presence of a cervical rib, post-surgical repair of coarctation of the aorta, congenital abnormalities like right aortic arch, and rare anatomical factors and congenital malformations [16].

There is a myriad of clinical presentations that can be observed in patients with SSS. One such finding is the difference in radial pulse (lowered or even absent) and systolic blood pressure (greater than 20mmHg) compared to contralateral upper extremity. Upper extremity ischemic symptoms can also be seen such as fatigue, exercise-related aching, coolness, and/or numbness of the involved upper extremity [15]. Neurological symptoms may also accompany SSS, including, presyncope, vertigo, headache, seizures, and confusion. It is thought that decreased blood flow from the basilar artery to the area of the brain responsible for balance and coordination is responsible for these symptoms [17].

A diagnosis of SSS requires both the characteristic clinical manifestations and a clinically correlated angiographic or duplex ultrasound finding of retrograde flow in the vertebral artery [18]. SSS can be classified into three grades based upon the hemodynamic instability seen in the ipsilateral vertebral artery [6].

- Grade I (pre-subclavian steal): reduced antegrade vertebral flow
- Grade II (intermittent/partial/latent): alternating flow - antegrade flow in the diastolic phase and retrograde flow in the systolic phase
- Grade III (permanent/advanced): permanent retrograde vertebral flow

In this case, the coarctation of the ARSA provided anatomical conditions for an uncommon right sided SSS. SSS tends to be less common on the right side, as only a small distance separates the bifurcation of the innominate artery and the branching of the vertebral artery [15]. ARSA present in this case likely preserved right carotid artery flow in the presence of right sided SSS and resulting in the right vertebral artery remaining an effective collateral blood flow pathway to the right upper extremity.

Surgical Implications: There is much surgical significance in regard to the ARSA and NRLN. Many complications can arise from routine procedures if the ARSA and NRLN are not identified and planned for preoperatively. One such case was reported in 2023 where an anterior discectomy and fusion of the C6-C7 vertebrae resulted in dysphonia [19]. Evidence of an ARSA was apparent on MRI preoperatively, and the decision to perform a right sided cervical approach as opposed to a left sided one likely resulted in damage of the NRLN [19].

In the case of the NRLN, the most significant complications can arise in patients undergoing a partial or total thyroidectomy. Without proper dissection and isolation of the recurrent laryngeal nerve, no transverse connection should be cut between vessel and larynx except the middle thyroid vein [20]. If the recurrent laryngeal nerve is not identified, there is substantial risk of damaging the motor and sensory innervation of the larynx [21].

Although rare, some patients with an ARSA can present with complications of dysphagia, cough, stridor, thorax pain or aneurysmal dilation, which can

necessitate surgical intervention [22]. In such cases, the ARSA is dissected from the posterior left arch of the aorta and retracted anteriorly to the esophagus and ultimately anastomosed with the right common carotid artery, all the while monitoring the location of the recurrent laryngeal nerve [22]. It is therefore imperative to meticulously identify and isolate the RRLN or NRLN in all surgical cases involving the neck and thorax of a patient identified with an ARSA.

CONCLUSION

This cadaveric case study presented an ARSA with proximal stenosis consistent with potential for SSS, and a NRLN. An ARSA often leads to a direct impact on the course of the recurrent laryngeal nerve creating the potential for the nerve to become non-recurrent. Abnormal neurovasculature must be identified preoperatively to avoid laceration of sensorimotor innervation to the larynx by the NRLN particularly during thyroidectomy and cervical spine procedures. Additionally, carotid anastomosis with the ARSA is required of patients presenting with dysphagia, respiratory symptoms, or aneurysmal dilation to restore circulation and relieve symptoms. Surgeons and clinicians must be vigilant in identification of rare anatomical variations, such as an ARSA, to provide the best overall care to their patients as failure to identify can lead to significant impact in surgical operations and patient well-being.

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