Rare case of a five-branched aortic arch exhibiting a retroesophageal right subclavian artery and an accessory left vertebral artery

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ABSTRACT

Head and neck vascular variations are common in humans, but often go undetected. They are generally asymptomatic. Awareness of such anatomical variations is clinically important for surgeons and interventional radiologists as they may pose risk for iatrogenic complications or even the potential for unanticipated fatalities. Although aortic arch variations are relatively common, branch variations are unanticipated fatalities.

As they may pose risk for iatrogenic complications or even the potential for unanticipated fatalities. Such knowledge is even more essential with the increasing use of carotid and vertebral artery stent placements and new therapeutic options for intracranial interventions (3,9,10).

Approximately 1 to 3% of human fetuses examined critically exhibit aortic arch (AA) variations (11). The most common form of AA variation is the aberrant subclavian artery (ASA; 0.1% to 2.5% incidence and more than 100 cases described in the literature (7,8,11-38). The incidence of an aberrant right subclavian artery (ARSA) is much higher and 80% of these are retroesophageal (RRSA). An aberrant left subclavian artery (ALSA) occurs in one in 1000 people (17,18,21). In these RRSA cases, the right brachiocephalic trunk (RBT) is absent and the following four arteries emerge as the AA branches: the right common carotid artery (RCCA), left common carotid artery (LCCA), the left subclavian artery (LSA), and the RRSA. In 13.7% of RRSA cases, the right vertebral artery (RVA) originates from the RCCA (19,32,36,39). Maiti et al. 2016 found that 80% of the 49 reported cases of RVA originating from the RCCA had an RRSA (38). Other vertebral artery (VA) variants are prevalent in both number and origin, but are documented infrequently in the literature (40-49). Unilateral duplication of the VA or an accessory VA (AVA) is considered a rare vascular variant as a fourth AA branch (0.295%-0.72%) (1,6,40-53).

We discovered during anatomical cadaveric dissection of a 76-year-old White male, provided by the Maryland State Anatomy Board, an additional branch. This individual exhibited an AA with five branches: RCCA, LCCA, LSA, ARSA (an RRSA), and an accessory left vertebral artery (ALVA) (Figure 1). The presence of a fifth AA branch accompanying the aforementioned variations, an ALVA, has not, based on these authors’ literature search, been described previously, and thus, may be a rare variant.

CASE REPORT

This 76-year-old White male (listed cause of death of acute kidney failure) presented with a five-branched AA. The branches included a RCCA, LCCA, LSA, ARSA (an RRSA), and an ALVA (Figures 2 and 3). A left vertebral artery (LVA) was present as the first branch off of the LSA. Both the RVA cadaveric dissection.

When aberrant subclavian arteries are described in adults, they frequently involve the right side, and are usually retroesophageal with an absent right brachiocephalic trunk. The following four branches of the aortic arch found most commonly then are: a retroesophageal right subclavian artery, left subclavian artery, and right and left common carotid arteries. During cadaveric dissection of a 76-year-old White male, we observed an additional fifth branch: an accessory left vertebral artery. A combination of five such aortic arch vessels may be rare.

Key Words: Retroesophageal right subclavian artery; Aberrant right subclavian artery; Accessory left vertebral artery; Duplicate origin of the left vertebral artery; Vertebral artery variation; Head and neck vascular variations
and ALVA entered the foramen transversarium at the C4 level, while the LVA entered at the C6 level. The RVA had a diameter of 3.5 mm. The LVA was hypoplastic, with a diameter of 1 mm. As the LVA traveled cranially and prior to merging with the ALVA, it gave off two small radiculomedullary (RMA) branches. The first RMA traveled medially and entered between the C5 and C6 vertebral bodies, and the second RMA entered between C4 and C5. The length of the LVA was 5.4 cm before uniting with the ALVA within the C4 foramen transversarium. As the ALVA ascended cranially, it exhibited stenosis with a diameter ranging between 4 and 5 mm. The circle of Willis also presented with several variations: the right posterior communicating artery (RPCA) was slightly hypoplastic while the left posterior communicating artery (LPCA) was markedly dilated, and the RVA had an area of stenosis just before it joined the ALVA to become the basilar artery (Figure 4). There were no variations detected with the cerebellar or basilar arteries.

Several of the RRSA branches were damaged during student dissection of the root of the neck, including the right costocervical trunk. Assessment of the branches by further careful dissection revealed that the right thyrocervical trunk (RTT) arose from a common trunk with the right internal mammary (thoracic) artery (RIMA). The RTT was the origin of a large number of the RRSA branches (Figure 5). These branches included the right transverse cervical artery (RTCA), right ascending cervical artery (RACA), and the right suprascapular artery (RSSA). The right inferior thyroid artery (RITA) was its own branch from the RRSA. The RVA originated from the RCCA instead of the RRSA. Figure 4 also highlights that the right recurrent laryngeal nerve (RRLN) did not loop around the RRSA.

The LSA branched slightly differently from the RRSA, along with other branching variations (Figure 6). As commonly seen, the LVA is the first branch off of the LSA. Here, the left inferior thyroid artery (LITA) was an independent branch, similar to the RRSA, but it originated from a common trunk with the left ascending cervical artery (LACA). Like the RRSA, the left thyrocervical trunk (LTT) arose from a common trunk with the left internal mammary (thoracic) artery (LIMA). The branches of the LTT included the left transverse cervical artery (LTCA) and the left suprascapular artery (LSSA). Only a small portion of the LSSA was present since it was damaged during student dissection of the root of the neck. However, the left costocervical trunk (LCT) was not damaged during student dissection.

**DISCUSSION**

**Aberrant right subclavian artery**

Aberrant right subclavian artery (ARSA) or arteria lusoria is a rare congenital vascular variation that is usually asymptomatic or symptoms appear later in life (7,12,13,17-22,24,27,29,31,35,37,54-56). Symptomatic patients usually exhibit dysphagia (termed dysphagia lusoria), dyspnea, cough, pneumonia, stridor, and/or thoracic pain. These symptoms are usually associated with compression of the adjacent structure (esophagus and/or trachea). Adults most typically present with dysphagia, while infants
to tracheal compression (7,8,11-14,16-19,21,23-25,27,30-33,35,37,54-56,59,62-66).

Other complications associated with an ARSA include atherosclerosis, abdominal aortic aneurysms (10%-20% of cases), stenosis, and dissections (23,24,65). ARSA also associates with other anatomical variations, such as a non-recurrent right inferior laryngeal nerve, a common origin of the CCAs (truncus bicaroticus) (20.0%-29% of cases), ipsilateral VA arising from the CCA, an anomalous point of entrance of the VA into the cervical spine, a replaced right or left VA, coarctation of the aorta, rightsided AA, and an aberrant right thoracic duct (8,13,14,16-19,30-33,36,57).

Most human embryos study develop six pairs of AAs that arise from the dextrocardia and migrate to the left heart. The LCCA is retracted so show the branches of the AA. The right fourth AA usually becomes the most proximal segment of the right subclavian artery (RSA). The distal part of the RSA forms from the union of a portion of right dorsal aorta and the seventh intersegmental artery. The portion of the right arch distal to the origin of the RSA regresses with the RCCA and RCA, thereby merging to form the BTA. The RSA is not derived from a pharyngeal arch artery; it forms from the left seventh intersegmental artery. As the development proceeds, differential growth shifts the origin of the LSA cranially, resulting in its final placement close to the origin of LCCA. The left fourth arch forms the AA that continues and descends on the left side of the spine as the descending aorta (7,11,15,18,21,33,35,38). In 64.9%-94.3% of cases, the AA has three great vessels as its branches from right to left: RBT, LCCA, and LSA. The RBT branches into the RSA and the RCCA. Such a pattern is considered typical (8,19,25,26,28,30,33,55).

In rare cases, the right fourth aortic arch involutes or disappears; an abnormal situation leaving the seventh intersegmental artery attached to the descending aorta. As a result, the RSA develops from the right seventh intersegmental artery and part of the right dorsal aorta caudal to the intersegmental artery (7,11,15,20,24,26,30,32,39,59,64,65). When the right sixth dorsal segmental artery persists, the RVA arises from the RCCA. In addition, persistent attachment of the first or second dorsal intersegmental arteries to the fourth left aortic arch, associates with the origin of the LVA from the aortic arch (7,11,15,20,24,26,30,32,39,59,64,65). Under these circumstances, the AA has four branches; the RCCA, LCCA, RSA, and the ARSA. The recurrent laryngeal nerve does not pass around the ARSA in any of these three path scenarios (67). If the regression occurs on the left arch, the result is an ALSA (11-14,21,27,37,57,64).

ARSA has been reported more frequently in females. The prevalence of ARSA rises exponentially to 26%-34% in individuals with Down syndrome (Trisomy 21) and other chromosomal defects, including microdeletion 22q11 (DiGeorge syndrome) and Trisomy 18 (Edward syndrome) (7,11,15-17,19,29,32,37,54,55,68-71). These are genetic conditions likely to survive into postnatal, and increasingly adult life. Such conditions frequently demonstrate vascular variations, particularly those of the heart, great vessels, and the AA.

**Accessory vertebral artery**

Unilateral duplicate origin of the VA or accessory vertebral artery (AVA) is considered rare, with a reported incidence of 0.295%-0.72% (1,6,40,50-53). In this individual, the left fifth intersegmental artery persisted. AVAs are equally frequent on both the right and left sides (72). This developmental variation associates with a dual origin and variable level of fusion with the VA in the neck (1,51,73-76). The AVA and VA can enter in the same foramen transversarium or in different foramina transversarium; usually the medial
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one enters at the higher level. Fusion of the AVA and VA occurs at the C5-C6 in 42.3% of cases, C4-C5 in 30.8%, C3-C4 in 15.4%, and C6-C7 in 11.5% (72,77). We found reports of 58 cases involving this vascular variant, with the first reported case in 1844 (2,6,10,40,46,51,52,73-75,77-92).

AVAs are usually asymptomatic clinically (1,3,4,6,28,53,78,88,93,94). This origin variation may alter cerebral hemodynamics, however, and lead to cerebral dysfunction. Such dysfunction may predispose a patient to cerebrovascular pathologies, such as aneurysm and dissection (2,3,6,9,75,81,85,95-100).

AVAs result from a developmental variant of the AA with the persistence of intersegmental arteries (5,101). For accessory RVAs (ARVs), originating from the RSA or BT, the right fourth or fifth or sixth (or less frequently the third) intersegmental artery persists. For the ALVA, the left fourth or fifth or sixth (less frequently the third) intersegmental artery persists (1,2,25,46,51,77). Additionally, the level at which the accessory VA enters the foramen transversarium indicates which intersegmental artery or arteries persist (6,9,51,82,85).

Circle of willis variations

The vascular variations we observed within the circle of Willis and cerebellum in this case are well-documented in the literature. Hypoplasia is one of the most frequent vascular anomalies in the circle of Willis, ranging from a 23%-27% incidence rate (102-105). Variation occurred in only 1.4% (108). The inferior thyroid artery originated directly from the thyrocervical trunk (37). In Takafuji and Sato 1991 study of 72 Japanese adults, only 3.5% exhibited a thyrocervical trunk that arose from a common trunk with the internal mammary (thoracic) artery (108,109). Its branches were the suprascapular artery and transverse cervical artery. In another study from the subclavian artery in 8% of cases in another study [37]. Variant origin of the RIMA occurred in 5% of cases examined with a frequency of 30% in LIMA in yet another study (110).

Another five-branched aortic arch description

Ma et al. described five vessels arising from the aortic arch. However, their unique case study reveals the following branches: RCCA, LCCA, left thyrocervical trunk (LTT), LSA, and RRSA (59). Our findings differ in that we found our case study to exhibit: RCCA, LCCA, LSA, RRSA, and an ALVA.

CONCLUSION

Aortic arch and subclavian branching variations are common and significant in both their origin and course of arteries. Many are asymptomatic but can have serious implications for clinical diagnosis, angiographies, interventional and surgical procedures in the head and neck. With the increasing use of a transradial approach for coronary angiography, due to its lower risk of associated access site related complications, ARSAs will be encountered more frequently. Awareness of the presence of ARSAs together with the presence of a right non-recurrent laryngeal nerve is clinically important when performing thyroid surgery. Awareness of an RRSA is significant, especially during right axillary, brachial, or radial angiographic approaches to the ascending thoracic aorta. The potential, but rare complication of a nearby arteriovenous fistula carries a 50% risk of rupture and fatality. In this case, we present the presence of the ALVA rather than a LVA originating from the AA as a genuine potential variation in this region. Its presentation with a hypoplastic LVA could have been missed easily had we not engaged in a continued thorough cadaveric dissection of this individual. Anatomists, radiologists, and surgeons need to recognize this possibility and thus, the importance of thorough dissection and imaging of this region.

ACKNOWLEDGEMENT

We would like to thank the family of our donor for their beneficent contribution. Without their generosity, this article would not have been possible. We would also like to thank Dr. Gary Wind for providing us an accurate anatomical rendering of the neurovasculature discussed in this article.

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