CASE REPORT

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An unusual case of three branches arising from the aortic arch: bicarotid trunk, left subclavian artery and aberrant right subclavian artery

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SUMMARY

The presence of an aberrant right subclavian artery originating as the last branch of the aortic arch is a rare variation. A single case of this aberrant vessel, together with a bicarotid trunk, was observed in an 80-year-old female during dissection. A small, atherosclerotic saccular aneurysm was present at the origin of the aberrant vessel. The aberrant right subclavian artery coursed posterior to the esophagus and trachea to reach the right upper limb. Surgeons operating on structures in the mediastinum, including the esophagus, should be aware of the existence of this variant vessel to avoid accidental injury.

Key words: Aberrant right subclavian artery – Retro-esophageal right subclavian artery – Bicarotid trunk **–** Aneurysm – Aortic arch – Anatomical variation

INTRODUCTION

Variation in the origin of branches arising from the aortic arch is common. The typical description is of three branches, the first being the brachiocephalic trunk, which divides and gives rise to the right subclavian artery and the right common carotid artery, the second branch is the left common carotid artery, while the final branch is the left subclavian artery (Natsis et al., 2017).

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An aberrant right subclavian artery (ARSA) is a variation in which, instead of being the first branch, this vessel arises as the last branch of the arch, or even from the descending aorta (Chadha and Chiti -Batelli, 2004). The artery must then travel from the left side of the body to the right upper limb, and is therefore required to navigate its way around structures of the mediastinum, such as the trachea and esophagus. The vessel passes between the esophagus and vertebral column in the majority of cases, although it may travel between the trachea and esophagus or, rarely, anteriorly to the trachea (Chadha and Chiti-Batelli, 2004; Natsis et al., 2017).

Aberrant right subclavian artery is a rare occurrence, and has been reported to have a prevalence of 0.2% to 13.3%, with a recent meta-analysis calculating a weighted pooled prevalence of 1.3% worldwide (Natsis et al., 2017; Polednak, 2017).

The presence of an aberrant right subclavian artery may be associated with a bicarotid trunk, in which the two common carotid arteries arise from a common origin from the aortic arch. The prevalence of a bicarotid trunk has been reported in up to 20% of individuals with an ARSA, although some studies have reported a lower prevalence of 0.8% (Polguz et al., 2014; Natsis et al., 2017). Occasionally, an aneurysm, previously described as a Kommerell's diverticulum, may be present at the origin of the aberrant right subclavian artery (Rogers et al., 2011). The right recurrent laryngeal nerve is typically non-recurrent in the presence of an aberrant right subclavian artery, instead coming off the Vagus nerve and travelling directly to the

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larynx (Natsis et al., 2017).

Although a rare variant, knowledge of the existence of an aberrant right subclavian artery is clinically relevant as it may produce symptoms that cannot otherwise be explained. Symptoms result from the compression of the trachea and esophagus, which may lead to dyspnea or dysphagia or even both, in up to 10% of patients with an ARSA (Polguz et al., 2014; Natsis et al., 2017).

CASE REPORT

During dissection of the aortic arch and its main branches in 85 cadavers (34 females and 51 males) in 2017 and 2018, a single case of aberrant right subclavian artery was discovered in an 80-year-old, formalin-embalmed female. The mediastinum was dissected to expose the heart and great vessels, with connective tissue and fascia overlying these structures removed in order to visualize the origins of the arteries. Careful dissection revealed three branches arising from the arch, of which a bicarotid trunk was the first branch, and the left subclavian artery, the second. The third branch was the right subclavian artery, which was situated distal and posterior to the left subclavian artery.

This vessel, originating as the last branch of the aortic arch instead of as the first branch, from the brachiocephalic trunk, traversed between the esophagus and vertebral column to reach the right upper limb, giving off its usual arterial branches, including the right vertebral artery (Fig. 1). The heart and great vessels were excised and removed from the body to inspect for the presence of an aneurysm at the origin of the aberrant vessel (Fig. 2).

A small saccular aneurysm was observed at the origin of the ARSA, which was found to contain atherosclerotic plaques. The course of the right recurrent laryngeal nerve could not be determined due to previous dissection by medical students. No medical history was available for the individual, and thus it is unknown whether the ARSA was symptomatic in this case. There were no other variations in the anatomy of the mediastinum and

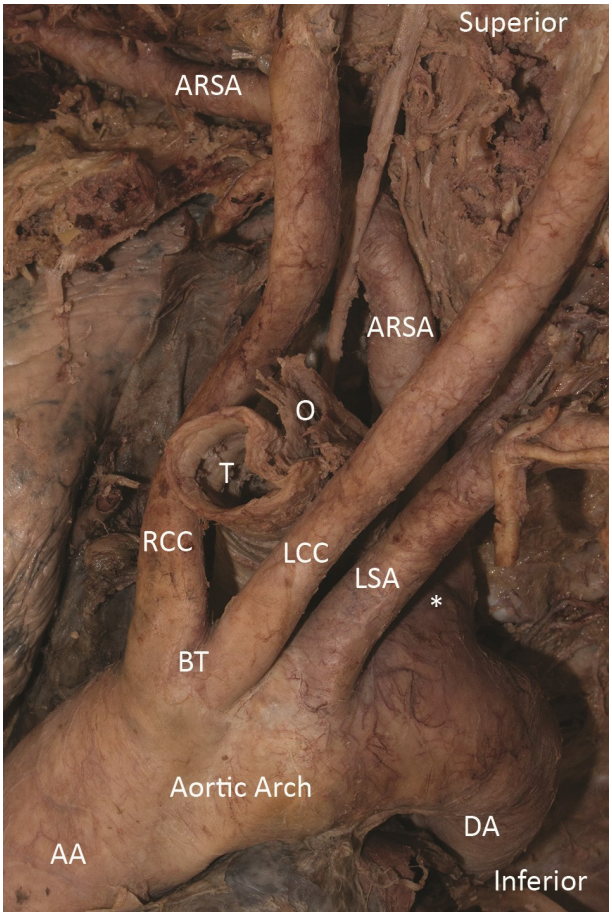

Fig 1. An aberrant right subclavian artery (ARSA) originating as the last branch of the aortic arch, in combination with a bicarotid trunk (BT) and aneurysm (*), as observed on dissection. The ARSA coursed behind the trachea (T) and esophagus (O) to reach the right upper limb. Abbreviations: AA ascending aorta; DA descending aorta; RCC right common carotid artery; LCC left common carotid artery; LSA left subclavian artery.

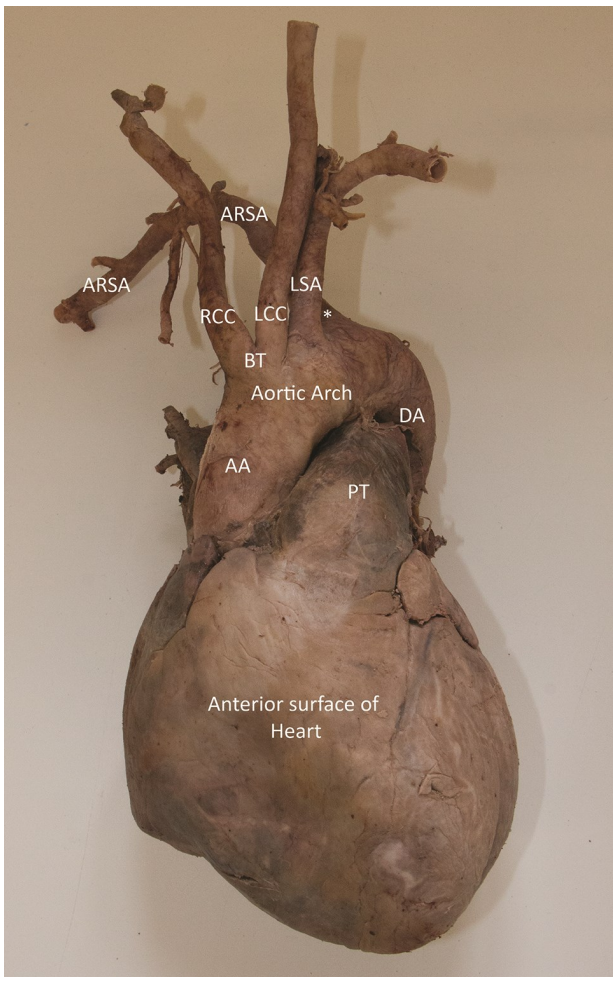

Fig 2. The origin (*) and course of the aberrant right subclavian artery (ARSA) as observed when the heart was removed. The small aneurysm was obscured by the origin of the left subclavian artery (LSA) Abbreviations: AA ascending aorta; PT pulmonary trunk; DA descending aorta; BT bicarotid trunk; RCC right common carotid artery; LCC left common carotid artery.

neck.

DISCUSSION

The development of an ARSA can be explained by the absence of the right fourth aortic arch. There are two dorsal aortas present during embryological development, with a left and a right arch, derived from the fourth pair of aortic arch arteries. The proximal part of the right subclavian artery originates from the right fourth arch and the distal part from the involution of the right dorsal aorta and right seventh intersegmental artery (Rogers et al., 2011; Scala et al., 2015; Natsis et al., 2017). The seventh intersegmental arteries originate from the descending aorta. If the fourth arch is absent, the seventh intersegmental artery remains affixed to the descending aorta or distal part of the aortic arch. In this situation, an aberrant right subclavian artery originating from the descending aorta or aortic arch results (Rogers et al., 2011; Natsis et al., 2017). The mechanism determining the course of the aberrant right subclavian artery relative to the trachea and esophagus is not well understood (Natsis et al., 2017).

The presence of a common origin of the left and right common carotid arteries reflect an alteration in the usual the development of these vessels. The common carotid arteries arise from the third pair of aortic arches, which originate from the aortic sac. As the base of the third pair of arches is formed by the sac (Moore and Torchia, 2016), a common origin of the left and right common carotid arteries may result if this part of the sac persists (Mustafa et al., 2017).

The presence of a bicarotid trunk occurring with an ARSA has been reported in the literature (Polguz et al., 2014; Natsis et al., 2017). Failure of the development of the fourth aortic arch together with persistence of the aortic sac may result in the coexistence of these two variant vessels. An aneurysm at the origin of the ARSA may be observed, although the prevalence varies widely from 1.7% to 60% (Polguz et al., 2014; Natsis et al., 2017). A bicarotid trunk and the presence of a small, atherosclerotic saccular aneurysm was observed in our individual with the ARSA. This variation has been previously observed in a patient at our institution, who presented with dysphagia and significant weight loss. An ARSA was eventually revealed by CT investigation after other tests and therapies failed to resolve the dysphagia (Rogers et al., 2011).

As the aberrant right subclavian artery passes behind the esophagus in most cases, dysphagia is the most common symptom (Natsis et al., 2017). Dyspnea is associated with an ARSA that passes between the trachea and esophagus, although it is also observed in infants in whom the ARSA is retro-esophageal in position, as the trachea is less rigid than in adults and therefore more likely to be compressed (Polguz et al., 2014). Other symptoms include stridor, cough, difficulty for eating, retrosternal pain, pulmonary infection, weight loss, stomach and back pain and right upper limb numbness (Natsis et al., 2017). “Arteria lusoria” refers to an ARSA that is diagnosed as the cause of dysphagia, while “dysphagia lusoria” describes dysphagia that presents as a result of the ARSA (Rogers et al., 2011). Dysphagia may, however, only develop with increasing age, as atherosclerotic plaques build up in either the vessel or an associated aneurysm, and result in the compression of the esophagus (Polguz et al., 2014). A bicarotid trunk associated with ARSA has also been implicated as the cause of dysphagia, as this large vessel anterior to the trachea and esophagus restricts the anterior movements of these structures (Rogers et al., 2011). Magnetic resonance angiography and computed tomography imaging are the best methods for diagnosing the presence of an ARSA (Natsis et al., 2017).

Knowledge of the possibility of the existence of an ARSA is important to surgeons operating in the mediastinum and neck regions. Failure to recognize this variation may result in fatal consequences. While performing a tracheostomy on a patient in 2004, Chadha and Chiti-Batelli observed a large artery overlying the trachea, which was identified as an aberrant right subclavian artery. Careful dissection and modification of the tracheostomy enabled these surgeons to avoid accidentally injuring the artery. Another patient with an ARSA and bicarotid trunk was not so fortunate (Satyapal et al., 2003). This patient had undergone emergency surgery for a stab wound superior to the sternum. Due to the injury, the anatomy of the aortic arch could not be visualized and what was thought to be the brachiocephalic trunk was clamped. However, the patient died postoperatively, and postmortem examination revealed the existence of an ARSA and a bicarotid trunk, which was mistaken for the brachiocephalic trunk, resulting in massive infarcts when clamped. ARSA may also be at risk of injury during surgery on the esophagus if not identified prior to procedures (Mahmodlou et al., 2014).

Several studies have shown an association between the presence of an aberrant right subclavian artery and certain conditions, such as congenital heart disease and Down syndrome. A meta- analysis suggests that the presence of an ARSA in fetuses, as detected by ultrasound, may be a significant risk marker of Down syndrome (Scala et al., 2015). Other risk markers can be assessed in fetuses with an ARSA and karyotyping performed if indicated.

Usually, no intervention is required for an ARSA unless there is an associated aneurysm or symptoms of esophageal or tracheal compression (Settembre et al., 2017). Repair of ARSA and reconstruction of blood flow to the right upper limb can be performed with a hybrid procedure of combined open surgery and endovascular techniques (Rogers et al., 2011; Polguz et al., 2014; Settembre et al., 2017). The origin of the ARSA from the aorta is closed, the retro-esophageal part of the vessel is removed and the remainder of the artery is transposed onto a suitable artery, such as the

right common carotid artery (Rogers et al., 2011).

Although a rare variation, the presence of an ARSA should be considered as a differential diagnosis for dysphagia if initial tests do not reveal any abnormalities and symptoms do not resolve with treatment.

CONCLUSION

An aberrant right subclavian artery originating from the aortic arch and passing behind the esophagus to reach the right upper limb is a rare finding during dissection. Other rare variations may coexist with ARSA, such as the presence of an aneurysm at the origin of the aberrant vessel or a bicarotid trunk, as observed in this case.

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ETHICS APPROVAL

Ethical approval for the use of human cadavers in research at our institution is approved under a blanket statement by the Human Research Ethics Committee, and thus no formal consent was required.

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