

ABNORMAL ORIGIN OF RIGHT SUBCLAVIAN ARTERY-EMBRYOLOGICAL BASIS AND CLINICAL SIGNIFICANCE

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ABSTRACT

BACKGROUND

Variations in the branching of the human aortic arch have clinical significance. 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. ARSA usually remains asymptomatic and is revealed incidentally either radiologically or during anatomy dissection. It is associated with several congenital cardiovascular anomalies and some chromosomal syndromes.

MATERIALS AND METHODS

Fifty cadavers were dissected and observed for aortic arch branching pattern variations during routine undergraduate dissection over a period of two years in Institute of Anatomy, Madurai Medical College, Madurai, Tamilnadu.

RESULTS

One case of aberrant right subclavian artery was noted. The right subclavian artery arose from the right lateral surface of proximal part of descending aorta distal to ligamentum arteriosum. It was seen to course upwards, obliquely towards the right, passing posterior to trachea and oesophagus. The right recurrent laryngeal nerve on arising from the right vagus ran transversely towards inferior pole of the right lateral lobe of thyroid gland and entered the larynx behind the cricothyroid joint. The course of right laryngeal nerve was short and non-recurrent.

CONCLUSION

Awareness of this anatomical variation will provide useful knowledge to anatomists, radiologists, cardiologists, vascular and thoracic surgeons and thereby prevent injury to aberrant right subclavian artery and right recurrent laryngeal nerve during radiological and surgical interventions.

KEYWORDS

Aberrant Right Subclavian Artery, Aneurysm, Arch of Aorta, Dysphagia, Non-Recurrent Laryngeal Nerve.

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BACKGROUND

Anomalies in the vascular anatomy of neck are clinically significant. The right subclavian artery normally arises from the brachiocephalic trunk, which stems from the arch of aorta. An Aberrant Right Subclavian Artery (ARSA) arises as a separate branch from the arch of aorta or from the descending aorta, distal to the origin of left subclavian artery.¹ ARSA also called arteria lusoria though rare is a common variant of aortic arch anomaly.² It usually arises from the aorta distal to the origin of left subclavian artery and shows a variable course to reach the right arm.²

The prevalence range of ARSA in general population ranges from 0.5% to 1.8%.³ ARSA is usually asymptomatic and revealed as an incidental finding during radiological interventions or cadaveric dissections. Rarely, it can cause dyspnoea, dysphagia and acute ischaemia to right upper limb.¹ The only clinical indicator of an ARSA is dysphagia on swallowing.⁴ Bayford described patient with dysphagia secondary to oesophageal compression by an ARSA and coined the term "dysphagia lusoria."¹

ARSA is associated with several congenital cardiovascular anomalies and chromosomal abnormalities. Recently, it has been suggested that the prenatal occurrence of this vascular anomaly is substantially increased in Down syndrome where it can be found in up to 19-36% of cases.² An NRLN is almost always associated with the aberrant subclavian artery due to embryological reasons. Thus, a diagnosis of ARSA, in theory, can be predictive of an NRLN.⁴

The aim of this study is to describe the presence of ARSA among 50 dissected cadavers, and to refer to its incidence, course, embryological basis and clinical significance according to literature reports. This information will provide

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useful knowledge to anatomists, radiologists, cardiologists, and vascular and thoracic surgeons. Awareness of this anatomical variation while performing radiological and surgical interventions will preserve aberrant right subclavian artery and non-recurrent laryngeal nerve and prevent devastating complications.

MATERIALS AND METHODS

The present study was conducted on 50 cadavers in Institute of Anatomy, Madurai Medical College, Madurai, Tamilnadu, over a period of four years during routine dissection of MBBS students.

During thorax dissection of superior mediastinum, arch of aorta was clearly delineated and its branches were followed, relations to trachea and oesophagus were also noted. Recurrent laryngeal nerves on both sides were identified and its course was noted.

RESULTS

Out of the 50 cadavers, abnormal origin of right subclavian artery was noted in a 60-year-old female cadaver. Three branches were seen to arise from the convexity of arch of aorta from right to left- right common carotid artery, left common carotid artery and left subclavian artery. Absence of brachiocephalic trunk was noted. The right subclavian artery showed variation in its origin and course. The right subclavian artery arose from the right lateral surface of proximal part of descending aorta distal to ligamentum arteriosum. It was seen to course upwards obliquely towards the right passing posterior to trachea and oesophagus. On reaching the superior mediastinum, the right subclavian artery was seen lying lateral to the right common carotid artery, but on a deeper plane. The right recurrent laryngeal nerve on arising from the right vagus ran transversely towards inferior pole of the right lateral lobe of thyroid gland and entered the larynx behind the cricothyroid joint. The course of right laryngeal nerve was short and non-recurrent. The left recurrent laryngeal nerve on the other hand looped typically around the aortic arch. The cardiac branches of both vagus nerves entered the cardiac plexus formation in the thorax.



Figure 1. Anterior View of Aortic Arch and its Branches; A- Arch of Aorta, B- Right Common Carotid Artery, C- Left Common Carotid Artery, D- Left Subclavian Artery, E- Right Subclavian Artery

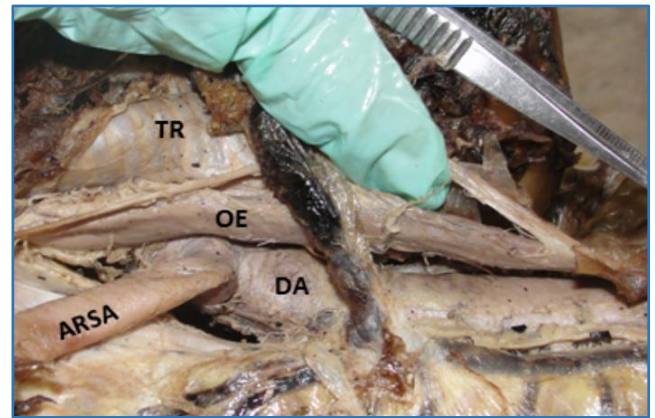


Figure 2. Right Lateral View of Posterior Mediastinum. TR - Trachea, OE - Oesophagus, DA - Descending Aorta, ARSA - Aberrant Right Subclavian Artery

DISCUSSION

Variations in the branching pattern of aortic arches are not uncommon. The arch of aorta normally gives rise to three branches- i) Brachiocephalic trunk, ii) Left common carotid artery, iii) Left subclavian artery. The brachiocephalic trunk branches into right subclavian artery and right common carotid artery.¹ ARSA also called arteria lusoria is a common variant of aortic arch anomaly. ARSA arises from aorta distal to the origin of left subclavian artery.⁵

The ARSA is usually described to arise from the dorsal margin of the aortic arch and in few cases from the descending aorta,⁶ but in the present report, the right subclavian artery arose from the right lateral surface of proximal part of descending aorta distal to ligamentum arteriosum. From its origin, ARSA is reported to cross the midline posterior to oesophagus to reach the right arm (80%), less commonly it passes between trachea and oesophagus (15%) or anterior to trachea (5%).¹ In the present case also, the anomalous artery was seen to course upwards obliquely towards the right passing posterior to trachea and oesophagus.

Variations in the right subclavian artery has been classified into ten types by Holzapfel in 1899 and the present anomaly belongs to type 5.⁷ According to the Adachi-Williams-Nakagawa classification, the present case belongs to type G. The incidence of the Adachi type G variation in Japanese adults has been reported to be from 0.15 to 1.6%.^{8,9,10} The prevalence range of ARSA in general population has been studied using different study materials and ranges from 0.5% to 1.8%.³ In Caucasian specimens, Williams et al found it once in 80 Caucasian cadavers.¹⁰

Fockens et al found 6 cases out of 1629 examined via endoscopic ultrasound.¹¹ Kelly found a single case in 223 patients undergoing upper gastrointestinal endoscopy for dysphagia.¹² Abaichan et al found 14 in 3730 patients undergoing transradial coronary angiography.¹³ Molz et al described a prevalence of 0.7% during autopsies.¹⁴ Hara et al studied the radiographic findings in 25 cases with prediagnosed aberrant artery on CT scans. On lateral radiographs, a posterior tracheal imprint was seen in 95%

of the patients.¹⁵ In 60% cases, the proximal part of ARSA is generally greater in diameter and may present a diverticulum. This saccular outpouching of aorta is referred to as "kommerell's diverticulum", it represents incomplete regression of primitive right aortic arch.¹⁶

Embryological Basis

Pharyngeal arches develop during 4th and 5th week of intrauterine life and are supplied by arteries from aortic sac. These arch arteries connect the embryonic aortic sac with paired dorsal aorta. Complex interplay of factors decide the formation and selective involution of aortic arches and its branches. Normally, the right subclavian artery develops from the distal fusion of a persistent right proximal dorsal aorta with the right seventh intersegmental artery. Aberrant right subclavian artery is the most common congenital aortic arch anomaly. ARSA is caused by abnormal obliteration of right fourth vascular arch and proximal right dorsal aorta and persistence of seventh intersegmental artery forming arteria lusoria.⁶ In addition and abnormal development of right fourth aortic arch results in the right inferior laryngeal nerve stemming directly from cervical part of the vagus nerve without having a recurrent course to reach the cricothyroid membrane.¹⁷

In the current study, right recurrent laryngeal nerve showed a non-recurrent course. Absence of right recurrent laryngeal nerve reported with a frequency of 0.3-1.6%.¹ The incidence of NRLN 0.21%-1.94% on the right side and 0-0.24% on the left side.⁴ ARSA usually remains asymptomatic, but rarely can cause dysphagia, dyspnoea, recurrent pulmonary infections, feeding difficulty and acute ischaemia to right upper limb. ARSA is revealed incidentally during anatomy dissection or radiologically.¹

The pressure applied by the ARSA onto the wall of the oesophagus might produce dysphagia. Tracheoesophageal compression by ARSA in children manifests in children mostly as respiratory symptoms, while in adults dysphagia is the principal symptom. Dysphagia lusoria in children can have a detrimental effect on dietary intake interrupting physical growth and cognitive development. Hence, it is imperative for early detection of ARSA in paediatric population.¹⁸

Aneurysm and dissection of an ARSA have been reported with an incidence of 3-8%.¹ Approximately, 90% of these aneurysms are due to atherosclerosis and 25% associated with aneurysms, elsewhere most notably aorta.¹⁶ Aneurysmal enlargement of ARSA is rare and presents as superior mediastinal mass.¹⁶ It is usually asymptomatic or causing dyspnoea, cough, swallowing difficulty, regurgitation, hoarseness, torticollis, vertigo, retrosternal pressure and pain referring to right arm. The walls of an ARSA are thinner and this carries high risk of spontaneous rupture or perforations of the oesophagus into an ARSA by foreign body or rupture of aneurysm into the oesophagus.¹ Stenosis or kinking of ARSA may cause unequal upper extremity blood pressure readings, right arm claudication, splinter haemorrhages or vertebrobasilar ischaemia.¹ ARSA-patients are at higher risk of iatrogenic injuries, while

performing imaging studies and catheter-based techniques for aortic arch and great vessels.¹⁹ Invasive procedures on the oesophagus such as thoracoscopic oesophagectomy can accidentally injure an ARSA.¹ Transient compression of ARSA during transesophageal echocardiography imaging can impair haemodynamic monitoring if radial arterial catheters are used on the affected side.¹ Due to embryologic reasons a NRLN is always associated with ARSA. Inadvertent damage to NRLN can result in hoarseness and pulmonary complications.⁴

Studies have found that of ARSA is associated with congenital cardiovascular anomalies and chromosomal abnormalities. 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.²⁰ Anatomic variations associated with ARSA include abnormal origin of right vertebral artery from aorta or from right common carotid artery, presence of common carotid trunk, right-sided thoracic duct and a non-recurrent laryngeal nerve.²¹

ARSA is often an accidental finding during investigative procedures for another cause is revealed incidentally during anatomy dissection.¹ ARSA can be diagnosed by plain chest x-ray, barium oesophagogram (oblique compression of the oesophagus at the level of third and fourth thoracic vertebrae), computed tomography images, magnetic resonance images and angiographies.¹⁹ CT or MRI is considered the gold standard for the diagnosis, since it helps to plan the operation and excludes other associated anomalies. It is difficult to preoperatively diagnose NRLN by any current imaging technique. Since, NRLN is always frequently associated with ARSA, preoperative identification of ARSA by chest enhanced CT scans enables diagnoses of NRLN and provide a wide image to reveal the complete course of ARSA. Recently, preoperative diagnosis of NRLN has attracted attention in thyroid and parathyroid surgeries.⁴

Treatment of ARSA depends upon the symptoms, age, comorbidity and concomitant vascular abnormalities of each patient.³ Surgical approach is indicated when ARSA is symptomatic or has evidence of aneurysm and varies from minimally-invasive procedures, endovascular techniques to more aggressive procedures.³ Though there is no standard surgical approach, right and left thoracotomies, cervical incision, median sternotomy, supraclavicular incision and combinations of these approaches have been used. In surgical treatment, anatomic reestablishment of orthograde flow into right subclavian artery avoids sacrificing direct blood flow to the right arm and possible complications thereof. Meticulous care should be taken during surgery to avoid unexpected arterial and nerve damage.⁴ Since progressive enlargement and rupture of this aneurysm is associated with high mortality rate, early elective resection is supported. Surgical repair would require a thoracotomy to control retroesophageal portion of ARSA and a supraclavicular incision for contralateral reconstruction.

Different operative procedures have been tried, including ligation of the aneurysm and anastomosing the right carotid artery to the distal right subclavian vessel. Newer procedures like transluminal endovascular grafting are less invasive and potentially less expensive with a lower risk than standard operative repair.¹⁶

Though ARSA is the most common aortic arch anomaly, it is an incidental finding in most of the cases. ARSA is most susceptible to iatrogenic injuries during surgical and radiological interventions. Detailed preoperative evaluation by chest enhanced CT scans might prevent damage to ARSA and NRLN. Knowledge about variations in the vascular anatomy of neck is crucial for general practitioners, radiologists, thoracic and vascular surgeons to prevent disastrous complications.

CONCLUSION

Vascular anomalies in the region of neck such as abnormal origin of right subclavian artery throws a significant clinical impact and should be borne in mind by radiologists, thoracic and vascular surgeons, interventional practitioners to avoid diagnostic pitfalls. Pre-operative identification and meticulous intraoperative dissection of ARSA would prevent disastrous vascular complications.

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