Aberrant Right Subclavian Artery. A Series of Case Reports and Discussion of Morphology and its Clinical and Surgical Implications

SUMMARY: Aberrant right subclavian artery (arteria lusoria) is a rare embryological abnormality but the most common among aortic arch vascular anomalies. It represents an anatomical variant of right subclavian artery originating as the last branch of aortic arch, passing then retroesophageal to the normal position. It is usually asymptomatic and is found mostly incidentally during imaging examinations. Symptoms are produced when the aberrant artery compresses the nearby structures and the most frequent symptoms are dysphagia and dyspnea. It may be associated with other vascular or heart abnormalities. We are presenting a series of case reports which presented an aberrant right subclavian artery alone or associated with other vascular abnormalities, diagnosed incidentally in adult patients while performing CT examinations for other reasons. Through a literature review we aim to discuss the clinical implications of this vascular anomaly, to point out the importance of being aware of it especially in patients with dysphagia or dyspnea or in patients who undergo operations in the thorax and neck or vascular surgery and endovascular procedures involving the aortic arch and its branches.

KEY WORDS: Arteria lusoria; Morphology; Clinical; Surgical; Implications.

INTRODUCTION

Aberrant right subclavian artery is an anatomical variation of the branches of aortic arch with an incidence varying from 0.2 to 3% of the population (Kieffer et al., 1994; De Araújo et al., 2015). Normally three branches arise from the convex aspect of the arch: the brachiocephalic trunk, left common carotid and left subclavian arteries (Standring, 2016). The right common carotid and subclavian arteries may arise separately, in which case the right subclavian artery often branches from the left end of the arch distal to the left subclavian artery. The vessel usually takes a retroesophageal path route to its usual site to the right arm, crossing the middle line of the body and may compress the esophagus producing dysphagia in some cases. The first description of an aberrant right subclavian artery was provided in 1735 by Hunauld, while dysphagia caused by it was described for the first time by Bayford in 1794 in a woman with a long history of dysphagia who in autopsy was found to have an aberrant right subclavian artery (Polguj et al., 2016). It was called dysphagia lusoria using the term “lusoria” from the Latin expression “lusus naturae,” which means “trick of nature” (de Oliveira Leite et al., 2017). In some cases, the lusoria artery arises from an aortic arch diverticulum at the proximal descending aorta which was first described by Kommerell in 1936 known as Kommerell diverticulum (Kommerell, 1936; Brown et al., 1993). According to Adachi–Williams’ classification the classic form of this variation is classified as type G, while more rare variations associated with lusoria artery are the origin of the left vertebral artery directly from the aortic arch (type CG), the common right and left carotid arteries arise from a unic trunk named bicaudal trunk (type H) or a right aortic arch with the left subclavian artery origin succeeding both carotid arteries and the right subclavian artery (type N) (Adachi, 1928; Williams et al., 1932).
CASE REPORTS

We are reporting a series of five case reports diagnosed incidentally with aberrant right subclavian artery. They didn’t complain of dysphagia or dyspnea and CT examination were performed for other reasons. Three patients resulted with *Arteria lusoria* type G, with retroesophageal course of aberrant artery, one patient with type CG (*Arteria lusoria* associated with the left vertebral artery originating directly from aortic arch) and one patient with type H (which represent *Arteria lusoria* associated with a bicarotid trunk) (Figs. 1 to 5).

DISCUSSION

Aberrant right subclavian artery is the most common anomaly of the aortic arch (Hanneman *et al*., 2017). The embryology of the branches of aortic arch evolves the branchial arches, the development of which begins by the second week of gestation and continues till the seventh week. There are six paired arches (numbered cranial-caudally) that connect the paired dorsal and ventral aorta (Kau *et al*., 2007). The first, second, and the fifth arches regress. The principal arches that form the aortic branches are the third, fourth and the sixth. The right subclavian artery has its origin in three sites: the fourth aortic arch which form the artery’s proximal portion, the segment from the right dorsal aorta between the forth aortic arch and the seventh right intersegmental artery and also the seventh right intersegmental artery (Kopp *et al*., 2007; Kellenberger, 2010). The aberrant origin of the right subclavian artery is caused by the involution of the right fourth vascular arch and proximal right dorsal aorta and the persistence of the seventh intersegmental artery originating from the proximal descending thoracic aorta, resulting in an abnormal artery course (Kopp *et al*., 2007).

The presence of Kommerell’s diverticulum varies from 15 % (Polguj *et al*., 2014) to 60 % of cases of Lusoria artery (Kommerell, 1936). Aneurysms are almost always found at or near the origin of the artery, and are suspected to result from degeneration of a diverticulum of Kommerell. These aneurysms should
always be treated because, even if asymptomatic, they can rupture, thrombose, or embolize (Kieffer et al., 1994).

The occurrence of *Arteria lusoria* in a systematic study was more common in female than male subjects and the most common symptoms produced by the compression of adjacent structures by an aberrant right subclavian artery were dysphagia, dyspnea, retrosternal pain, cough, and weight loss (Polguj et al., 2014).

According to a large bibliography search, the symptoms of *Arteria lusoria* compression have been found to be present only in 7–10% of adult patients with the anomaly. So, the anomaly is clinically silent in 90–93% of cases (Delap et al., 2000). When symptomatic, the aberrant right subclavian artery most often produces dysphagia, usually to solids without any difficulty in swallowing fluids from esophageal compression and is more frequent in older patients due to increased rigidity of the oesophagus itself or the vessel wall (Ulger et al., 2004; Myers et al., 2010; Reynolds et al., 2015). In infants, the trachea is compressible; therefore, the typical signs and symptoms of compression by lusoria artery are mainly respiratory, such as wheezing, stridor, recurrent pneumonia, and cyanosis (Derbel et al., 2012). Treatment for dysphagia lusoria varies

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**Fig. 3.** The aberrant right subclavian artery marked with white arrow passing retro esophageal (type G). Patient 3 (female).

**Fig. 4.** *Arteria lusoria* associated with left vertebral artery originating from aortic arch (type CG). Patient 4 (female, age 22). First row. Red arrow shows right vertebral artery originating from right subclavian artery (white arrow), left vertebral artery (yellow arrow) is not originated from left subclavian artery (blue arrow). Second row: left vertebral artery (yellow arrow) originating from arch of aorta; right aberrant subclavian artery originating from arch of aorta passing retroesophageal (white arrow) Third row: right subclavian aberrant artery (white arrow).
depending on the severity of the symptoms. Dietary modifications are recommended in patients with mild to moderate symptoms while vascular reconstruction is necessary for patients with severe symptoms (Janssen et al., 2000).

Another important symptom that may be present with a stenotic Arteria lusoria is upper extremities blood pressure difference, claudication, and Raynaud’s syndrome of the right hand (Tuleja et al., 2019). Arteria lusoria has been reported to be associated with other embryological anomalies like congenital heart anomalies most common of which are Tetralogy of Fallot, ventricular septal defect or atrioventricular canal defect (Myers et al., 2010). It is strongly associated with Down syndrome, and has also been associated with an aberrant right thoracic duct (Epstein & Debord., 2002).

Arteria lusoria may be associated with a right non-recurrent laryngeal nerve. Instead of recurring from the chest, it will pass directly from the vagus nerve at the level of the larynx to the neck (Nagayama et al., 1994). It is important to know in cervical surgery, as this nerve will not follow its usual course and is at risk of damage.

This aberrant vessel also has surgical significance, because of its spatial relations to many structures, and it can be damaged during surgical procedures. Awareness of vascular variations of aortic arch and its branches is very important to surgeons during surgical procedures in the thorax and neck in order to achieve desired objectives and to avoid major complications during vascular surgery (Myers et al., 2010; Pop et al., 2012; Lacout et al., 2012).

Therefore, surgeons who perform vascular surgery involving the aortic arch and its branches or endovascular procedures for example stenting of both stenotic and occlusive lesions of the supra aortic trunks or cardiologist who perform transradial coronarography should be warned of Lusoria artery as the most frequent anomaly of aortic arch encountered.

CONCLUSION

Aberrant right subclavian artery is a rare embryological abnormality. It is usually asymptomatic but may produce symptoms like dyspnea or dysphagia and should be taken into consideration during differential diagnoses. Proper knowledge of anatomic and morphologic variations of the arch of aorta and especially of the aberrant right subclavian artery which is the most frequent among them, is imperative in the diagnostic and surgical procedures in the thorax and neck.

**RESUMEN:** La arteria subclavia derecha aberrante (Arteria lusoria) es una anomalía embriológica rara, pero la más común entre las anomalías vasculares del arco aórtico. Representa una variante anatómica de la arteria subclavia derecha que se origina como la última rama del arco aórtico, pasando luego retroesofágicamente a la posición normal. Por lo general, esta anomalía es asintomática y se encuentra principalmente de manera incidental durante los exámenes de imagen. Los síntomas se producen cuando la arteria aberrante comprime las estructuras cercanas y los síntomas más frecuentes son la disfagia y la disnea. Puede estar asociado con otras anomalías vasculares o cardíacas. Presentamos una serie de informes de casos en los que se presentó una arteria subclavia derecha aberrante única o asociada a otras anomalías vasculares, diagnosticada incidentalmente en pacientes adultos durante la realización de TC por otros motivos. A través de una revisión bibliográfica pretendemos discutir las implicaciones clínicas de esta anomalía vascular, señalar la importancia de conocerla especialmente en pacientes con disfagia o disnea o en pacientes sometidos a operaciones de tórax y cuello o cirugía vascular y procedimientos endovasculares, involucrando el arco aórtico y sus ramas.

**PALABRAS CLAVE:** Arteria lusoria; Morfología; Implicaciones clínicas, quirúrgicas.

**REFERENCES**


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