Int. J. Morphol.,
404):1123-1127, 2022.

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

Arteria Subclavia Derecha Aberrante, una Serie de Informes de Casos y Discusión de la Morfología y sus Implicaciones Clínicas y Quirúrgicas

Marjeta Tanka[1]; Anila Kristo[2]; Nikollaq Leka[2] & Artan Kristo[3]

TANKA, M.; KRISTO, A.; LEKA, N. & KRISTO, A. Aberrant right subclavian artery. A series of case reports and discussion of morphology and its clinical and surgical implications. *Int. J. Morphol., 40(4)*:1123-1127, 2022.

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 bicarotid 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).

[1] Service of Imagery. University Hospital Center "Mother Tereza" Tirana, Albania.
[2] Department of Morphology, Faculty of Medicine. University of Medicine, Tirana, Albania.
[3] Service of Cardiology. University Hospital Center «Mother Tereza», Tirana, Albania.

Received: 2022-02-11 Accepted: 2022-05-12

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).

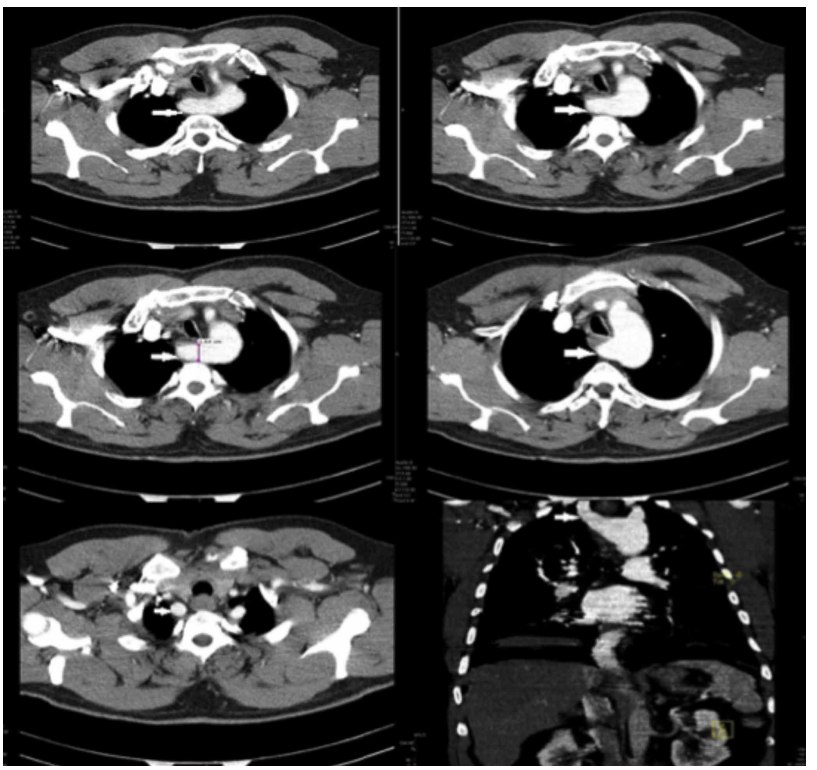

Fig.1. White arrow shows right subclavian aberrant artery (type G). Patient 1 (male, age 54).

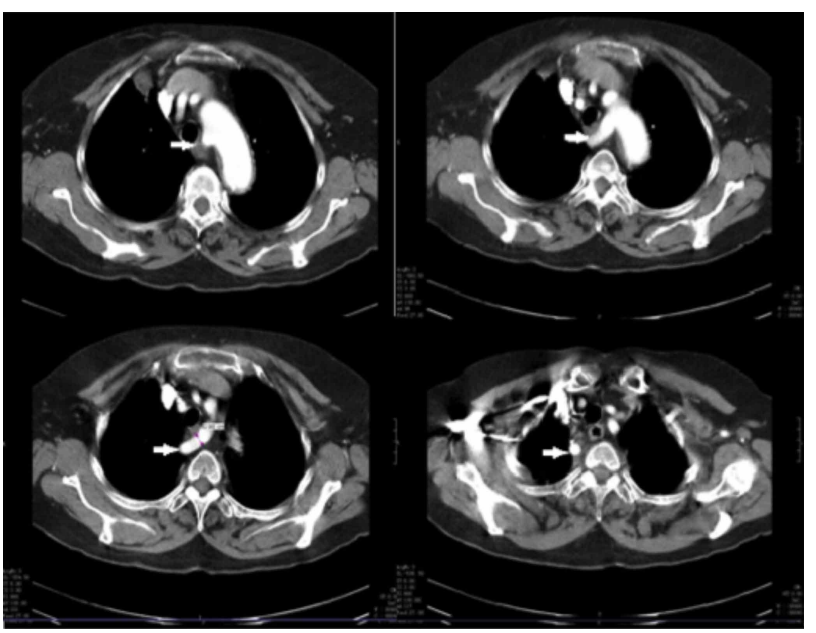

Fig. 2. White arrows show right subclavian aberrant artery (type G). Patient 2 (female, age 74).

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

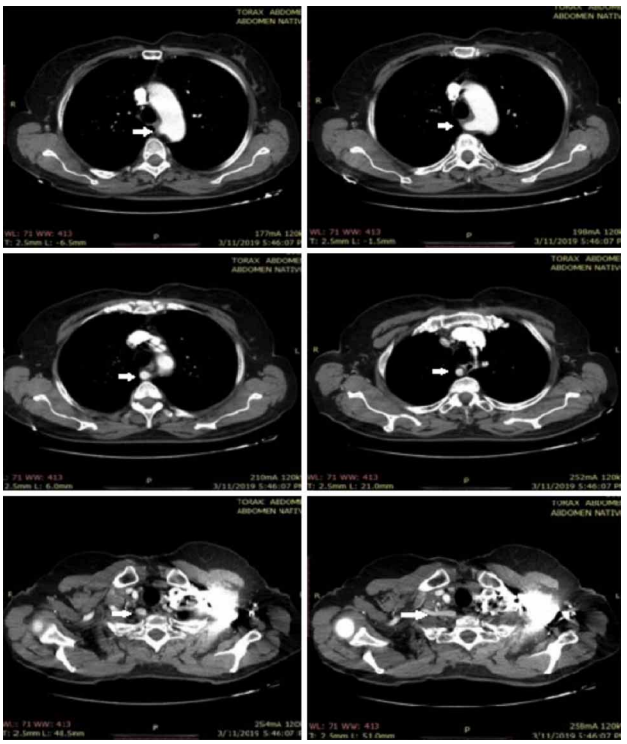

Fig. 3. The aberrant right subclavian artery marked with white arrow passing retro esophageal (type G). Patient 3 (female).

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

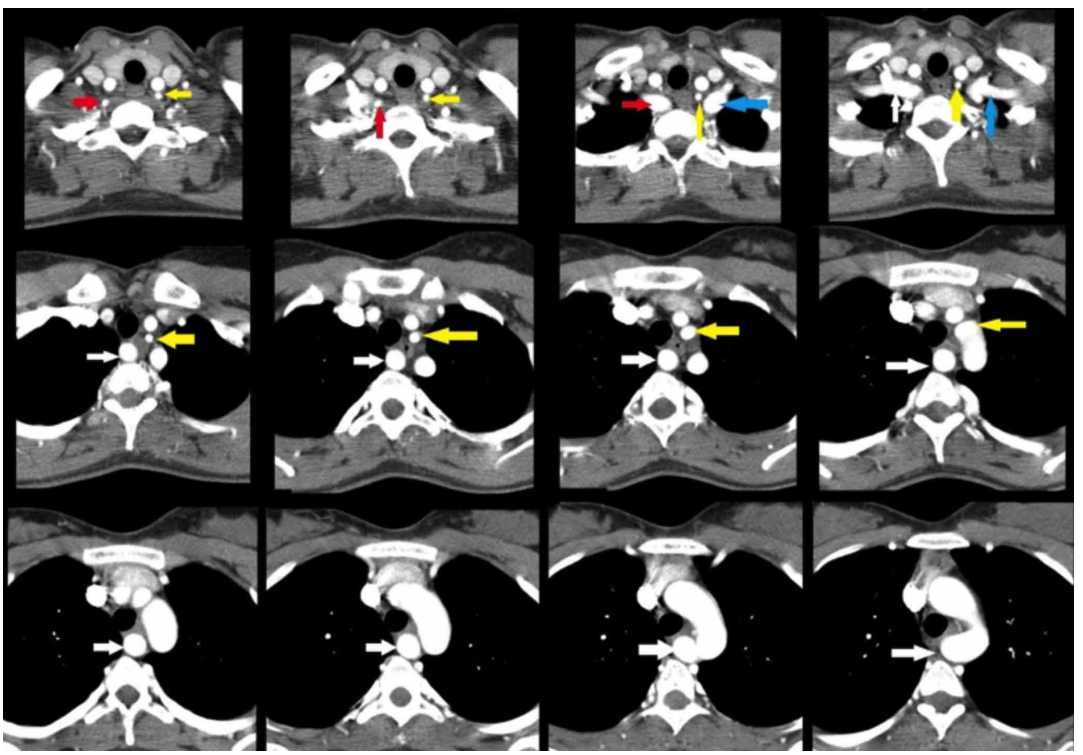

Fig. 4. *Arteria lusoria* associated with left vertebral artery originating from aortic arch (type CG). Patient 4 (female, age 22). First row. Red arrow shows right vertebral artery originating from right subclavian artery (white arrow), left vertebral artery (yellow arrow) is not originated from left 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).

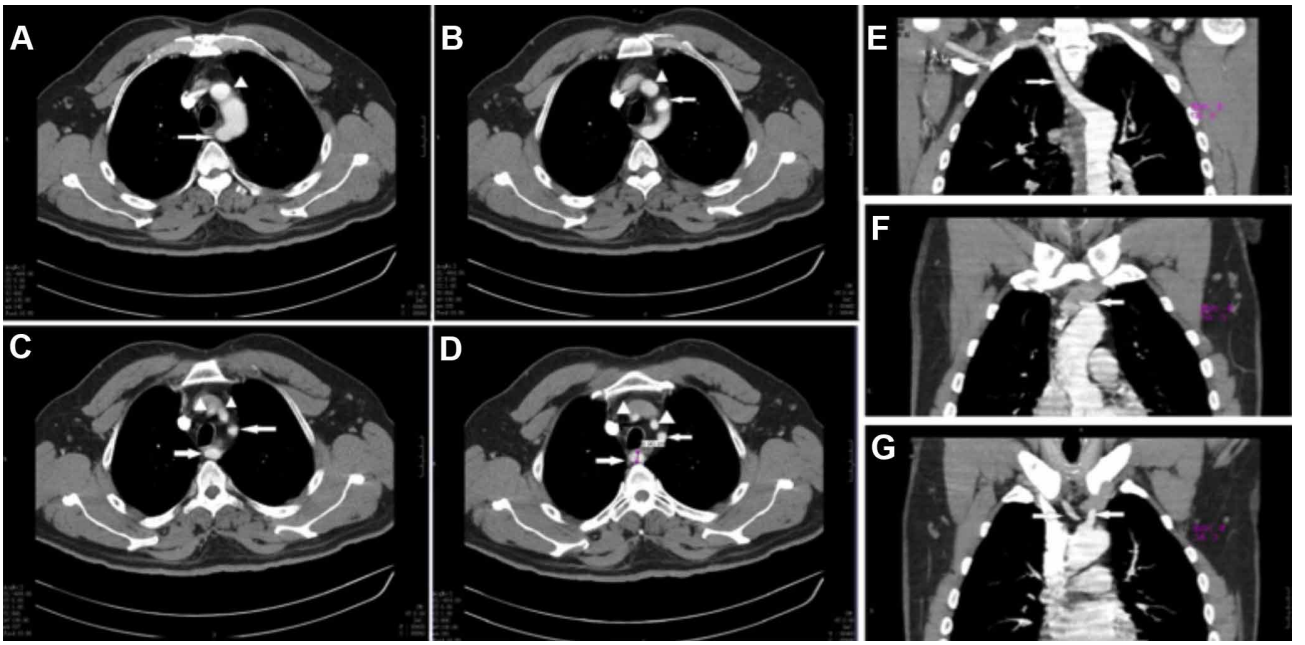

Fig. 5. *Arteria lusoria* associated with bicarotid trunk (type H). Patient 5 (female, age 43). A. arrow-right subclavian artery; arrowhead-bicarotid trunk. B. arrow-left subclavian artery; arrowhead-bicarotid trunk. C. arrows-right and left subclavian arteries; arrowheads-right and left common carotid arteries. D. arrows-right and left subclavian arteries; arrowheads-right and left common carotid arteries. E. arrow-right subclavian artery. F. arrow –bicarotid trunk. G. arrows-right and left common carotid arteries

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 awarned 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.

TANKA, M.; KRISTO, A.; LEKA, N. & KRISTO, A. Arteria subclavia derecha aberrante, una serie de informes de casos y discusión de la morfología y sus implicaciones clínicas y quirúrgicas. *Int. J. Morphol., 40(4)*:1123-1127, 2022.

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

Adachi, B. *Das Arterien System Der Japaner*. Kyoto, Verlag der Kaiserlich-Japanischen Universitat. Kenyusha Press, 1928. Vol. 1. pp.29-41.

Brown, D. L.; Chapman, W. C.; Edwards, W. H.; Coltharp, W. H. & Stoney, W. S. Dysphagia lusoria: aberrant right subclavian artery with a Kommerell's diverticulum. *Am. Surg., 59(9)*:582-6, 1993.

De Araújo, G.; Junqueira Bizzi, J. W.; Muller, J. & Cavazzola, L. T. "Dysphagia lusoria" - Right subclavian retroesophageal artery causing intermitent esophageal compression and eventual dysphagia - A case report and literature review. *Int. J. Surg. Case Rep., 10*:32-4, 2015.

de Oliveira Leite, T. F.; Pires, L. A. S.; Cisne, R.; Babinski, M. A. & Chagas, C. A. A. Clinical discussion of the *Arteria lusoria*: a case report. *J. Vasc. Bras., 16(4)*:339-42, 2017.

Delap, T. G.; Jones, S. E. & Johnson, D. R. Aneurysm of an aberrant right subclavian artery presenting as dysphagia lusoria. *Ann. Otol. Rhinol. Laryngol., 109(2)*:231-4, 2000.

Derbel, B.; Saaidi, A.; Kasraoui, R.; Chaouch, N.; Aouini, F.; Ben Romdhane, N. & Manaa, J. Aberrant right subclavian artery or *Arteria lusoria*: a rare cause of dyspnea in children. *Ann. Vasc. Surg., 26(3)*:419.e1-4, 2012.

Epstein, D. A. & Debord, J. R. Abnormalities associated with aberrant right subclavian arteries-a case report. *Vasc. Endovascular Surg., 36(4)*:297-303, 2002.

Hanneman, K.; Newman, B. & Chan, F. Congenital variants and anomalies of the aortic arch. *Radiographics, 37(1)*:32-51, 2017.

Janssen, M.; Baggen, M. G.; Veen, H. F.; Smout, A. J.; Bekkers, J. A.; Jonkman, J. G. & Ouwendijk, R. J. Dysphagia lusoria: clinical aspects, manometric findings, diagnosis, and therapy. *Am. J. Gastroenterol., 95(6)*:1411-6, 2000.

Kau, T.; Sinzig, M.; Gasser, J.; Lesnik, G.; Rabitsch, E.; Celedin, S.; Eicher, W.; Illiasch, H. & Hausegger, K. A. development and anomalies. *Semin. Intervent. Radiol., 24(2)*:141-52, 2007.

Kellenberger, C. J. Aortic arch malformations. *Pediatr. Radiol., 40(6)*:876-84, 2010

Kieffer, E.; Bahnini, A. & Koskas, F. Aberrant subclavian artery: surgical treatment in thirty-three adult patients. *J. Vasc. Surg., 19(1)*:100-9, 1994.

Kommerell, B. F. Verlagerung des Oesophagus durch eine abnorm verlaufende Arteria subclavia dextra (*Arteria lusoria*). *Forstschr. Geb. Roetgenstr., 54*:590-5, 1936.

Kopp, R.; Wizgall, I.; Kreuzer, E.; Meimarakis, G.; Weidenhagen, R.; Kühnl, A.; Conrad, C.; Jauch, K. W. & Lauterjung, L. Surgical and endovascular treatment of symptomatic aberrant right subclavian artery (*Arteria lusoria*). *Vascular, 15(2)*:84-91, 2007.

Lacout, A.; Khalil, A.; Figl, A.; Liloku, R. & Marcy, P. Y. Vertebral *Arteria lusoria*: a life-threatening condition for oesophageal surgery. *Surg. Radiol. Anat., 34(4)*:381-3, 2012.

Myers, P. O.; Fasel, J. H.; Kalangos, A. & Gailloud, P. *Arteria lusoria*: developmental anatomy, clinical, radiological and surgical aspects. *Ann. Cardiol. Angeiol. (Paris), 59(3)*:147-54, 2010.

Nagayama, I.; Okabe, Y.; Katoh, H. & Furukawa, M. Importance of preoperative recognition of the nonrecurrent laryngeal nerve. *J. Laryngol. Otol., 108(5)*:417-9, 1994.

Polguj, M.; Chrzanowski, L.; Kasprzak, J. D.; Stefanczyk, L.; Topol, M. & Majos, A. The aberrant right subclavian artery (*Arteria lusoria*): the morphological and clinical aspects of one of the most important variations--a systematic study of 141 reports. *ScientificWorldJournal, 2014*:292734, 2014.

Polguj, M.; Stefan´czyk, L. & Topol, M. *The Epidemiological, Morphological, and Clinical Aspects of the Aberrant Right Subclavian Artery (Arteria lusoria)*. In: Kasenga, F. (Ed.). Epidemiology of Communicable and Non-Communicable Diseases - Attributes of Lifestyle and Nature on Humankind. London, IntechOpen, 2016.

Pop, D.; Venissac, N.; Nadeemy, A. S.; Schneck, A. S.; Aze, O. & Mouroux, J. Lesson to be learned: beware of lusoria artery during transhiatal esophagectomy. *Ann. Thorac. Surg., 94(3)*:1010-1, 2012.

Reynolds, I.; McGarry, J. & Mullett, H. Aberrant right retroesophageal subclavian artery causing esophageal compression. *Clin. Case Rep., 3(10)*:897-8, 2015.

Standring, S. *Gray's Anatomy. The Anatomical Basis of Clinical Practice*. 41st ed. New York, Elsevier, 2016.

Tuleja, A.; Baumgartner, I. & Schindewolf, M. Claudication caused by stenosis of *Arteria lusoria*-case report and review of literature. *Clin. Med. Insights Case Rep., 12*:1179547619842187, 2019.

Ulger, Z.; Ozyürek, A. R.; Levent, E.; Gürses, D. & Parlar, A. *Arteria lusoria* as a cause of dysphagia. *Acta Cardiol., 59(4)*:445-7, 2004.

Williams, G. D.; Aff, H. M.; Schmeckebier, M.; Edmonds, H. W. & Graul, E. G. Variations in the arrangement of the branches arising from the aortic arch in American whites and negroes. *Anat. Rec., 54(2)*:247-51, 1932.

Corresponding author:
Anila Kristo
Department of Morphology
Faculty of Medicine
University of Medicine
Tirana
ALBANIA

E-mail: anilashukaus@yahoo.com