SHORT REPORT

Anomalies associated with aberrant right subclavian arteries. A case report

Elena Félix Domínguez, Blanca Mompeó Corredera

Departamento de Morfología, Universidad de Las Palmas de Gran Canaria, Gran Canaria, Spain

SUMMARY

Anomalies in the aortic arch are a consequence of disorders in the development of the double primitive aortic arch system.

We report a case of variation in the great vessels of the aortic arch, with an aberrant right subclavian artery being observed during a routine dissection. This variation was associated with a tight trachea in its distal end and a right lung devoid of the horizontal fissure, with a lack of tissue in the anterior segment of the superior lobe and in the lateral and medial segments of the middle lobe. The two common carotid arteries arose from a common trunk and the right recurrent laryngeal nerve was absent. On the basis of the literature, we review the incidence of the anatomical variation, its embryological explanation, and its clinical consequences.

Key words: Lusoria artery – Aberrant right subclavian artery – Anomalies in the aortic arch

INTRODUCTION

The structure of the human body is variable both externally and internally. Some of the

internal variations can produce symptomatology which may be interpreted as pathologic processes or it could entail surgical risks if they are not suspected.

Anatomical variations are frequent and it is necessary to be acquainted with them in order to establish a differential diagnosis when facing any types of symptoms. Dissection rooms are propitious places to observe the existence of human anatomical variations, the associations among them, and the clinical consequences of their presence.

The lusoria artery is an aberrant right subclavian artery, and is the last great vessel of the aortic arch and usually travels between the oesophagus and the vertebral column. The presence of anomalous arterial trunk may sometimes be overlooked or may be observed accidentally with diagnostic image techniques. Patients with this variation complain of compressive symptoms, particularly dysphagia, due to the posterior compression of the oesophagus (Del Val et al., 1997; Kollar et al., 1987; Aubertin et al., 1996; Jansen et al., 2000; Brown et al., 1993). This variation has also been described in relation to respiratory symptomatology as a consequence of traqueobronchial compression (Bove et al., 2001; Donelly et al., 2002). In addition, patients may present distal embolization (Werber et

Correspondence to:

Submitted: February 23, 2009 Accepted: April 22, 2009

Blanca Mompeó Corredera. Departamento de Morfología, ULPGC, Dr. Pasteur s/n, 35016 Las Palmas de Gran Canaria, España. Phone: 928453414; Fax: 928453420. E-mail: bmompeo@dmor.ulpgc.es

al., 1983) and aneurysm rupture (Steward et al., 1964) and the variation has been considered to be responsible for traumatic aortic rupture, so it has been considered that the presence of an aberrant right subclavian artery may be of potential risk for the patient (Schneider et al., 2007). Here we report the case of a Lusoria artery accompanied by other structural modifications.

CASE REPORT

In a routine dissection of the cadaver of a 70-year-old woman, we observed an aortic arch whose first branch was a common trunk from which both primitive carotid arteries arose; these arteries followed an oblique upwards and outside direction (Fig. 1). At a second site, the left subclavian artery arose from the arch and the right subclavian artery was the last branch (Fig. 2a, b). This artery ran towards the upper limb behind the oesophagus.

The laryngeal recurrent left nerve was observed around the aortic arch, posterior to the origin of the last trunk. The laryngeal recurrent right nerve could not be located.

The trachea was 9 cm long and consisted of 15 rings. A narrow part was seen in the antero-left side of its distal end. The external diameter of the superior traqueal rings was 23 mm and the inferior ones 18 mm.

The right lung lacked the smaller fissure and the pulmonary mass was scarce in the ante-

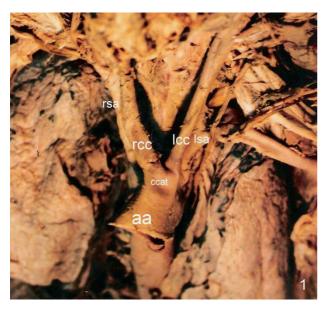


Fig. 1. Thorax. Brachiocephalic artery is absent. A common trunk for both common carotid arteries is the first branch in the aortic arch. aa: aortic arch; ccat: common carotid artery trunk; rcc: right common carotid artery; lcc: left common carotid artery; rsa: right subclavian artery; lsa: left subclavian artery.

rior segments of the superior lobe as well as in both segments of the middle lobe (Fig. 3).

Later dissection of the right bronchial tree revealed a distribution without variations.

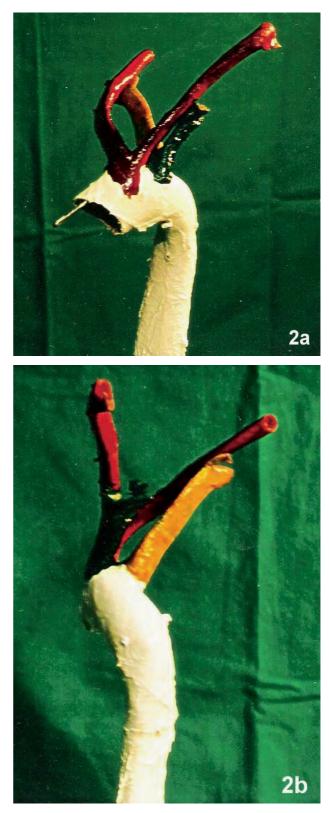


Fig. 2. Prepared aortic arch of the reported case and disposition of the vessels, 2a (anterior view), 2b (posterior view). Red: Common carotid trunk for both carotid arteries; green: left subclavian artery; yellow: right subclavian artery; white: aortic arch and descending aorta.

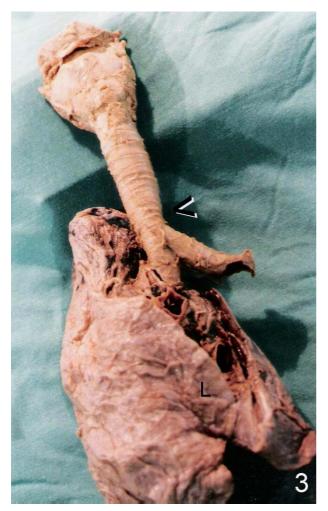


Fig. 3. Trachea and right lung. The trachea shows tightness in its distal end (arrow). There is no horizontal fissure in the right lung. The anterior segment of the superior lobe and the lateral and medial segments of the middle lobe become a lingular structure (L).

DISCUSSION

An aberrant right subclavian artery was described for the first time by Hunald in 1735. The reported incidence is 0.3-2% in autopsy studies, without differences between the sexes (De Luca et al., 2000). Aberrant right subclavian arteries have a retroesophagal location in 80% of the patients; they are located between the trachea and the oesophagus in 15%, and are anterior to both structures in 5% of cases.

An association of the anatomical variation with dysphagia was reported by Byford in 1787 (Miller et al, 1992). Although the presence of this variation may not be symptomatic, the aberrant right subclavian artery has been related to dysphagia (dysphagia lusoria) as a consequence of its frequent retroesophageal location (Del Val et al., 1997; Kollar et al., 1987; Aubertin et al., 1996; Jansen et al., 2000; Brown et al., 1993). This was the location in our case. The presence of dysphagia in adults with the anatomical variation is usually associated with atherosclerotic changes and aneurysms in the artery (Mok et al., 1979).

The presence of an aberrant right subclavian artery has also been related to respiratory symptomatology due to tracheobronchial compression (Bove et al., 2001; Donelly et al., 2002). Although respiratory alterations of vascular origin are rare, they can cause important distress and respiratory symptoms, fundamentally in children, since these have a tracheal wall that is less rigid than in adults (Kollar et al., 1987).

In our case, we observed the association of the lusoria artery with a trachea that showed a tightening at its distal end. Its lower rings had a diameter 20% smaller than the upper rings. The tightening could be the reason for the scant development of the pulmonary tissue in the anterior segment of the superior lobe and the lateral and medial segments of the middle lobe in the right lung, due to a decrease in air flow toward the corresponding segmental bronchi.

In the case reported here, the aberrant right subclavian artery was associated with a common origin of the carotid arteries from the aortic arch. The coincidence of both anomalies has a reported incidence of 29% in the patients with retroesophageal subclavian arteries (Klinkhamer, 1966).

The tracheal compression observed in our case, could have been a consequence of traction exercised by the right subclavian artery, with displacement of the posterior segment of the arch and descending aorta towards the middle line and the consecutive pressure of the common trunk of the common carotid arteries and aortic arch on the distal part of the trachea.

It has been shown that respiratory alterations occur in 50 % of paediatric patients having an aortic left arch with an aberrant right subclavian artery (Donelly et al., 2002). However, their diagnosis, with an unspecific respiratory symptomatology, may be late, with the consequent production of tracheobronchial malacia (Bove et al., 2001). The modification found in the case reported here featured the anatomical basis of a possible respiratory alteration secondary to the presence of the lusoria artery.

The presence of a non-recurrent laryngeal nerve is related to anomalies in the development of the aortic arch and of the supraaortic trunks. The reported incidence is 0.54% in the right side and 0.07% in the left side (Henry et al., 1985).

This association occurs due to the interrelationship between both structures during embryonic development. Thus, the formation of a right aberrant subclavian artery occurs during the sixth-eighth week of prenatal development and arises as a consequence of the involution of the fourth aortic right arch and of the dorsal aorta, cranial to the seventh intersegmental artery.

As a result of this abnormal regression, the right subclavian artery is formed from the seventh intersegmental artery and the distal portion of the dorsal right aorta. As the embryo grows, the origin of the right subclavian artery moves up to the origin of the left subclavian artery and eventually both locate very close together.

Although the primary reason for modifications in the disposition of the great vessels is not well known, they may be a consequence of the possible role of the cells originating in the neural crest during embryological development (Bergwerff et al., 1999).

The inferior laryngeal nerves bow around the sixth pair of the aortic arches to go pass the larynx during its growth.

On the right side, when the degeneration of the distal portion of the sixth aortic arch takes place under normal conditions the recurrent laryngeal nerve moves up, describing a curve around the proximal portion of the right subclavian artery derived from the fourth arch. Additionally, the left recurrent laryngeal nerve usually describes a curve around the ductus arterioso; if the fourth right arch is absent the nerve follows an atypical course.

Damage to the inferior laryngeal nerve is one of the main surgical risks in thyroid and parathyroid excision (Pisanu et al., 2002).

Since the presence of the nervous anomaly is in all the cases associated with the vascular anomaly (Henry et al., 1985), the finding of a right subclavian artery must alert the physician to the presence of an inferior laryngeal nerve of atypical location.

In sum, an aberrant right subclavian artery is considered an anatomical variation in itself, without pathological consequences. However, the variation may represent a potential risk for the patient, especially if the anomaly is not suspected. This variation should be considered when a patient exhibits dysphagia or symptoms of tracheobronchial compression. Moreover, its accidental finding should alert the clinician about the presence of a nonrecurrent laryngeal nerve in order to prevent laryngeal nerve damage in surgical interventions in the neck (Watanabe et al., 2001; Marchesi et al., 2000). References

- AUBERTIN JM, ISENI MC, TORDJMAN T, BLOCH F, PETITE JP (1995). Dysphagia Lusoria. Physiopathological discussion apropos of a clinical case. *Gastroenterol Clin Biol*, 19: 629-632.
- BERGWERFF M, DE RUITER MC, HALL S, POELMANN RE, GITTENBERG DE GROOT AC (1999). Unique vascular morphology of the fourth aortic arches: possible implications for pathogenesis of type B aortic arch interruption and anomalous right subclavian artery. *Cardiovasc Res*, 44: 185-196.
- BOVE T, DEMANET H, CASIMIR G, VIART P, GOLDSTEIN JP, DEUVAERT FE (2001). Tracheobronchial compression of vascular origin. Review of experience in infants and children. J Cardiovasc Surg, 42: 663-666.
- BROWN DL, CHAPMAN WC, EDWARDS WH, COLTHARP WH, STONEY WS (1993). Dysphagia Lusoria: aberrant right subclavian artery with Kommrell's diverticulum. *Am Surg*, 59: 582-586.
- DE LUCA L, BERGMANN JJ, TYTGAT GN, FOCKENS P (2000). EUS imaging of the artery lusoria: case series and review. *Gastrointest Endosc*, 52: 670-673.
- DEL VAL A, TALENS A, BELLOCH V, HERNÁNDEZ E (1977). Dysphagia lusoria caused by an accessory aberrant right subclavian artery. *Rev Esp Enferm Dig*, 89: 336-337.
- DONELLY LE, FLECK RJ, PACHARN P, ZIEGLER MA, FRICKE BL, COTTON RT (2002). Aberrant subclavian arteries: cross-sectional imaging findings in infants and children referred for evaluation of extrinsic airway compression. *AJR AM J Roentgenol*, 178: 1269-1274.
- HENRY JF, AUDIFFRET J, PLAN M (1985). The nonrecurrent inferior laryngeal nerve. Apropos of 19 cases including 2 on the left side. *J Chris*, 122: 391-397.
- JANSSEN M, BAGGEN MG, VEEN HF, SMOUT AJ, BEKKERS JA, JONKMAN JG, OUWENDIJK FJ (2000). Dysphagia Lusoria: clinical aspects, manometric findings, diagnosis and therapy. *Am J Gastroenterol*, 95: 1411-1416.
- KLINKHAMER AC (1966). Aberrant right subclavian artery: Clinical and roentgenological aspects. *Am J Roentgenol*, 97: 438.
- MARCHESI M, BIFFONI M, FALOCI C, NOBILI BENEDETTI R, NOTARI P, MARIOTTI F, CRESTI R (2000). The inferior nonrecurrent laryngeal nerve: a report of seven cases observed since 1987. *G Chir*, 21: 25-28.
- MILLER JM, MILLER KS (1992). A note on the historical aspects of dysphagia lusoria. *Am Surg*, 58: 502-503.
- Мок СК, CHEUNG KL, KONG SM (1979). Translocating the aberrant right subclavian artery in dysphagia lusoria. *Br J Surg*, 66: 113-116.
- PISANU A, PILI S, UCCHEDDU A (2002). Non-recurrent inferior laryngeal nerve. *Chir Ital*, 54: 7-14.
- STEWART JR, KINCAID OW, EDWARDS JE (1964). An Atlas of vascular rings and related malformation of the aortic arch system. Charles C. Thomas, Springfield, IL, pp 3-17, 80-129.
- WATANABE A, KAWABORI S, OSANI H, TANIGUCHI M, HOSOKAWA M (2001). Preoperative computed tomography diagnosis of non-recurrent inferior laryngeal nerve. *Laryngoscope*, 111: 1756-1759.
- WERBER J, RAMILO JL, LONDON R, HARRIS VJ (1983). Unilateral absence of a pulmonary artery. *Chest*, 84: 729-732.