Tortuosity of the internal carotid artery cervical course: case reports and literature review

Fazan, VPS.[1,2]*, Ribeiro, RA.[1], Oliveira, MAS.[1], Caetano, AG.[1] and Rodrigues Filho, OA.[1]

[1]Department of Biological Sciences, Human Anatomy Discipline, Federal University of Triângulo Mineiro, Uberaba, MG, Brazil

[2]Department of Surgery and Anatomy, School of Medicine of Ribeirão Preto, University of São Paulo, Av. Bandeirantes 3900, CEP 14049-900, Ribeirão Preto, SP, Brazil

*E-mail: vpsfazan@yahoo.com.br; vpsfazan@gmail.com

Abstract

Variations in the course of the internal carotid arteries (ICA) are reported in the literature as coiling, looping, kinking or tortuosities of the vessels. Nevertheless, the definitions between these variants are confusing. Also, the clinical relevance of morphological anomalies of extracranial ICA is a matter of debate because of up to date the natural history of kinking, coiling and tortuosities of this artery is not well known. However, some authors consider that these conditions are burdened with disabling, even fatal neurological complications. Also, variations of the ICA cervical course may lead to direct contact of the artery with the pharyngeal wall, being of great clinical relevance due to the large number of routine procedures performed in this region. In the present study, we describe two cases of ICA tortuosities and review the current literature regarding the causes, symptoms and clinical significances of the variations of the cervical ICA course. Tortuosity of the cervical ICA is not a rare condition and they can easily be mistaken clinically for an aneurysm, a tumor or an abscess and subsequently injured during an attempted biopsy or excision. Thus, regardless the controversy of its causes (congenital or acquired) it should be included in the differential diagnosis of cervical soft tissue widening. Also, they should be taken into consideration on the diagnostic procedures for ischemic transitory attacks and/or stroke.

Keywords: internal carotid artery, carotid kinking, carotid coiling, anatomical variation, vascular surgery.

1 Introduction

The origin and course of the carotid arterial system in the superior mediastinum and neck are remarkably constant. Although variations are rare, they can have important implications in certain clinical and/or surgical problems. Variations in the course of the internal carotid arteries (ICA) are reported in the literature as coiling, looping, kinking or tortuosities of the vessels. Nevertheless, the definitions between these variants are confusing. Most of these terms describe only the visual impressions created by two-dimensional angiografic films (WITZ and LEHMANN, 2001) while it seems that the real basic problem is an excessive length of the artery (ILLUMINATI, CALIÓ, PAPASPYROPOULOS et al., 2003; PANCERA, RIBUL, PRESCIUTTINI et al., 2000; SZÉKELY and CSÉCSEI, 2001).

Based on systematic postmortem statistics and extensive angiographic investigations, the incidence of variations in the cervical course of the ICA is reported to lie between 10 and 40% of the population (LA BARBERA, LA MARCA, MARTINO et al., 2006; WEIBEL and FIELDS, 1965), the variations usually being normally bilateral (CAIRNEY, 1924; METZ, MURRAY-LESLIE, BANNISTER et al., 1961; PAULSEN, TILLMANN, CHRISTOFIDES, 2000). The clinical relevance of morphological anomalies of extracranial ICA is a matter of debate because of up to date the natural history of kinking, coiling and tortuosities of this artery is not well known (LA BARBERA, LA MARCA, MARTINO et al., 2006). Some authors consider these conditions due to a benign angiopathy (PERDUE, BARRECA, SMITH et al., 1975) while others suppose that they are burdened with disabling, even fatal neurological complications (DERRICK and SMITH, 1962; HUEMER, EMMINGER, TRATTNIG et al., 1998; ILLUMINATI, CALIÓ, PAPASPYROPOULOS et al., 2003; ÖZBEK, YETKIN, ÖZELÇI et al., 2007; QUATTLEBAUM Jr, WADE and WHIDDON, 1973; SCHUMACHER, SCHAFIG, KEHRL et al., 1998; SPADONI, NERI, LUTI et al., 2003; TRACKLER and MIKULICICH, 1974). Moreover, it is well known that variations of the ICA cervical course may lead to direct contact of the artery with the pharyngeal wall, being of great clinical relevance due to the large number of routine procedures performed in this region (PAULSEN, TILLMANN, CHRISTOFIDES et al., 2000).

In the present study, we describe two cases of ICA tortuosities and review the current literature regarding the causes, symptoms and clinical significances of the variations of the cervical ICA course.

2 Case report

Fifty male embalmed human cadavers (fixed in 10% formaldehyde solution) were included in the present study. The necks have been partially dissected by medical students during the previous years and further dissections were performed by the authors, with the aid of a D. F. Vasconcellos M-90 surgical microscope. Bilateral evaluation of the carotid arterial system was done in each cadaver and two of them presented a cervical ICA with an unilateral tortuosity described as follows. None of these cadavers had previous history of hypertension and both arteries evaluated had no macroscopic signs of atheromatous/occlusive disease. With the aid of an electronic digital caliper (range of 0-300 mm, resolution 0.01 mm, Gehaka, SP, Brazil), the ICA diameter was measured as previously described (FAZAN, BORGES, SILVA et al., 2004; RIBEIRO, RIBEIRO, RODRIGUES FILHO et al., 2006), both at the superior and inferior ends of the tortuosity as was the diameter of the ICA at the tip of the artery folding.

Case 1: A 50 years old male cadaver presented a right ICA with a postero-lateral folding (Figure 1a), behind the internal jugular vein, crossing over the middle scalene muscle. This folding was at the level of the second and third cervical vertebrae (C2-C3). The beginning of the folding was located at 1.3 cm superior to the common carotid artery bifurcation. The diameter of the ICA was 1.0 cm either at the inferior or the superior ends of the folding. The diameter of the artery at the tip of the folding was 0.7 cm.

Case 2: A 53 years old male cadaver presented a left ICA with a posterior folding (Figure 1b), crossing over the middle scalene muscle and extending over the levator scapulae muscle. This folding was at the level of the atlas and the axis vertebrae (C1-C2). The beginning of the folding was located at 2.9 cm superior to the common carotid artery bifurcation. The diameter of the ICA was 1.2 cm at the inferior and 0.5 cm at the superior ends of the folding. The diameter of the artery at the tip of the folding was 0.6 cm.

3 Discussion

It is well known that the intracranial portions of the ICA are highly tortuous, while its cervical course is almost straight. There are two points of fixation at the cervical portion of the ICA: at the bifurcation of the common carotid artery and at its entry into the temporal bone. If the vessel is longer than the distance between these two points, curvatures and loops develop (SZÉKELY and CSÉCSEI, 2001). Anomalies of the carotid system are known to pose difficulty to surgeons of this area. Several authors consider these variations as congenital anomalies of the aortic arch arteries (CAIRNEY, 1924; JÄCKEL, 1997; PAULSEN, TILLMANN, CHRISTOFIDES et al., 2000; PRIOR, JOHNSON, JONES et al., 1997) while others consider it

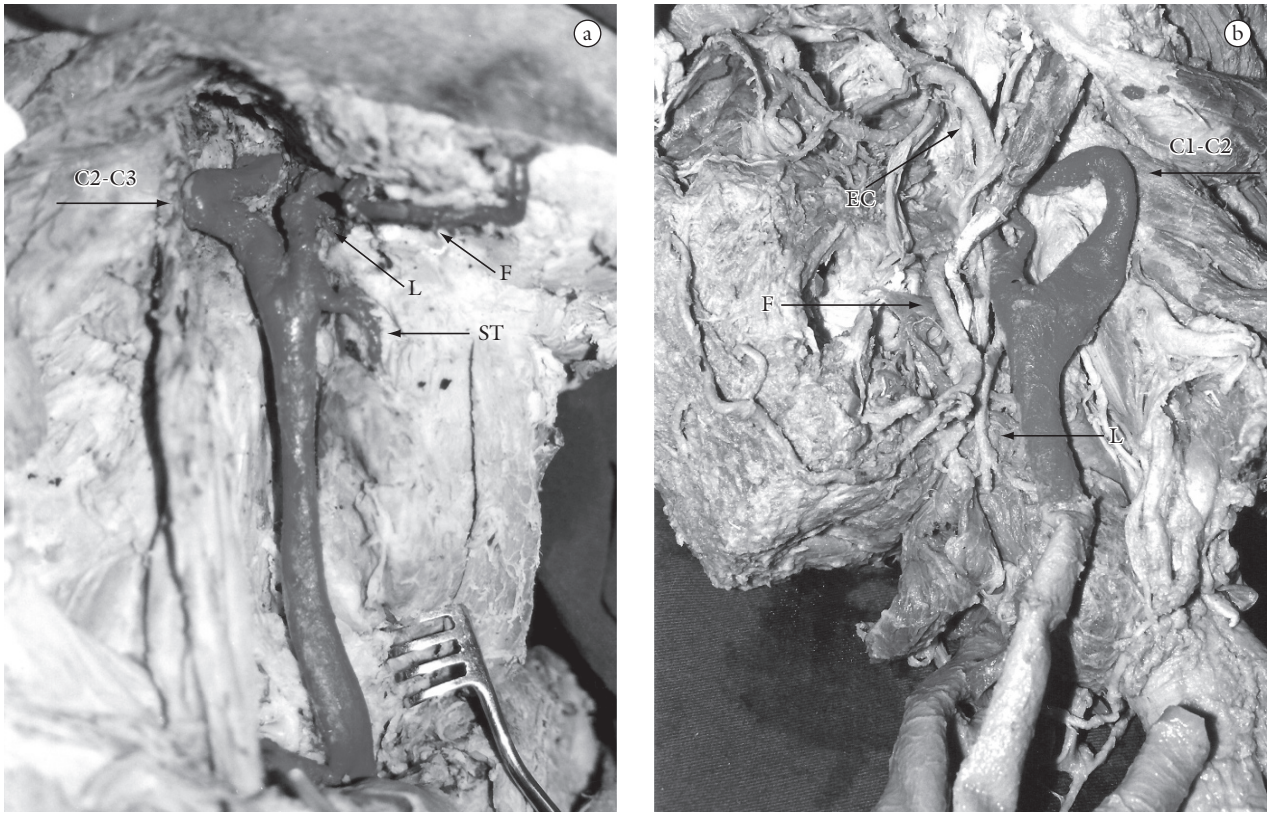

Figure 1. a) Folding of the right internal carotid artery at the level of second and third cervical vertebrae (C2-C3). Note that this folding has a very sharp posterior angle, crossing above the middle scalene muscle. F = facial artery, L = lingual artery, ST = superior thyroid artery; and b) Folding of the left carotid artery at the level of atlas and axis (C1-C2) vertebrae. This folding has a less sharp posterior angle but it is longer, crossing over the middle scalene muscle and reaching the levator scapulae muscle. F = facial artery, L = lingual artery, EC = external carotid artery. In both figures, the carotid arteries were colored with red latex, in order to enhance the contrast with other anatomical structures.

being caused by artheriosclerosis, fibromuscular hyperplasia (PAULSEN, TILLMANN, CHRISTOFIDES et al., 2000; SZÉKELY and CSÉCSEI, 2001), degenerative lesions (JÄCKEL, 1997; LA BARBERA, LA MARCA, MARTINO et al., 2006; QUATTLEBAUM Jr, WADE, WHIDDON, 1973), hypertension (OLIVIERO, COCOZZA, PICANO et al., 1997; PANCERA, RIBUL, PRESCIUTTINI et al., 2000) and/or aging (Del CORSO, MORUZZO, CONTE et al., 1998; ILLUMINATI, CALIÓ, PAPASPYROPOULOS et al., 2003; PANCERA, RIBUL, PRESCIUTTINI et al., 2000; PAULSEN, TILLMANN, CHRISTOFIDES et al., 2000). This controversy is still an unsolved issue. In the present cases, no signs of arterial lesions were found, either in the carotid system or in other arteries that could lead us to an acquired condition causing the tortuosities. On the other hand, both cases show the tortuosity in only one side of the neck. The congenital cause for the tortuosities can be explained in terms of the embryological development of the branchial arteries and is reinforced by the fact that cervical ICA tortuosities were found in children (CAIRNEY, 1924; HUEMER, EMMINGER, TRATTNIG et al., 1998; KELLY, 1925; LE BRET, PINEAU, FOLLIGUET et al., 2000; PERDUE, BARRECA, SMITH et al., 1975). One interesting point that should be taken into account is that if there is an abnormal descent of the large blood vessels and the heart to the mediastinal space during the embryo development, which leads to the elongation and straightening of the arteries, than the tortuosities would be more likely bilateral. However, despite few descriptions of bilateral cases (DUNCAN, SHER and PENCHARZ, 2002; KELLY, 1925; LE BRET, PINEAU, FOLLIGUET et al., 2000) most of the literature descriptions are of unilateral cases, like the ones we present here and there are also descriptions of the right side being more affected than the left (CAIRNEY, 1924; KELLY, 1925).

The variety of the terms used in order to define a carotid artery tortuosity adds to the confusion about its origin and incidence. Coiling, looping, S- or C-shaped elongation and kinking are only some of the terms most commonly used to define these variants. From all these, kinking of the ICA has the most reliable definition being an angulation of one or more segments of the artery (HUEMER, EMMINGER, TRATTNIG et al., 1998), forming an acute angle (ILLUMINATI, CALIÓ, PAPASPYROPOULOS et al., 2003; SPADONI, NERI, LUTI et al., 2003). Due to this controversy, we believe that tortuosity is a more general term that can be used to define any of the variants of the ICA cervical path.

It has been reported that clinical symptoms of the ICA tortuosity seldom occur and only a small percentage of the cases are diagnosed (DEL CORSO, MORUZZO, CONTE et al., 1998; KELLY, 1925; SZÉKELY and CSÉCSEI, 2001). Nevertheless, when these tortuosities are in direct contact with the pharyngeal wall, patients can present symptoms such as difficulties of swallowing and speech or even sensations of a foreign body in the area of the pharynx (OKAMI, ONUKI, ISHIDA et al., 2001; PAULSEN, TILLMANN, CHRISTOFIDES et al., 2000). The haemodynamic consequences of the ICA tortuosities remain controversial but significant associations with hemispheric (HUEMER, EMMINGER, TRATTNIG et al., 1998) and focal neurological (SPADONI, NERI, LUTI et al., 2003) symptoms have been reported. With the increasing use of arteriography, many patients have been found to have twisted or kinked carotid arteries, and the relation of this finding to cerebral insufficiency has become apparent (DERRICK and SMITH, 1962; METZ, MURRAY-LESLIE, BANNISTER et al., 1961). Our study shows a reduction of the diameter of the ICA at the tortuosity level in both cases, which can be an indication that the tortuosity might cause a reduction of flow within the vessel (TRACKLER and MIKULICICH, 1974) that might be exacerbated by progressive head rotation up to the point that causes complete cessation of flow (ILLUMINATI, CALIÓ, PAPASPYROPOULOS et al., 2003), thus depending on the position of the head (LA BARBERA, LA MARCA, MARTINO et al., 2006; QUATTLEBAUM Jr, WADE and WHIDDON, 1973). Tourtuous extracranial ICA needs no treatment as long as the patient does not have a cerebrovascular ischemic sign (OKAMI, ONUKI, ISHIDA et al., 2001) or a neurological complaint (RIBEIRO, RIBEIRO, RODRIGUES FILHO et al., 2006).

4 Conclusions

Tortuosity of the cervical ICA is not a rare condition (DERRICK and SMITH, 1962; KELLY, 1925; OKAMI, ONUKI, ISHIDA et al., 2001; PERDUE, BARRECA, SMITH et al., 1975) and they can easily be mistaken clinically for an aneurysm, a tumor or an abscess and subsequently injured during an attempted biopsy or excision. Thus, regardless the controversy of its causes (congenital or acquired) it should be included in the differential diagnosis of cervical soft tissue widening. Also, they should be taken into consideration on the diagnostic procedures for ischemic transitory attacks and/or stroke.

Acknowledgements: We thank the late Mr. Arnaldo Geraldino, Federal University of Triângulo Mineiro, Department of Biological Sciences, for excellent technical support while the study was carried out. Grant sponsor: FAPESP (Fundação de Amparo à Pesquisa do Estado de São Paulo); Grant numbers: 06/03200-7 and 06/06362-8; Grant sponsor: CNPq (Conselho Nacional de Pesquisa e Tecnologia); Grant number: 303802/2006-5.

References

CAIRNEY, J. Tortuosity of the cervical segment of the internal carotid artery. *J. Anat.* 1924, vol. 59, no. 1, p. 87-96.

DEL CORSO, L., MORUZZO, D., CONTE, B. et al. Tortuosity, kinking, and coiling of the carotid artery: Expression of atherosclerosis or aging? *Angiology.* 1998, vol. 49, no. 5, p. 361-371.

DERRICK, JR. and SMITH, T. Carotid kinking as a cause of cerebral insufficiency. *Circulation,* 1962, vol. 25, p. 894-853.

DUNCAN, IC., SHER, BJ. and PENCHARZ, M. Kinking of bilateral internal carotid arteries that mimics posttraumatic radiographic prevertebral soft-tissue widening. *Am. J. Roentgenol.* 2002, vol. 179, no. 5, p. 1352.

FAZAN, VPS., BORGES, CT., SILVA, JH. et al. Superficial palmar arch: an arterial diameter study. *J. Anat.* 2004, vol. 204, no. 4, p. 307-311.

HUEMER, M., EMMINGER, W., TRATTNIG, S. et al. Kinking and stenosis of the carotid artery associated with homolateral

ischaemic brain infarction in a patient treated with cyclosporine A. *Eur. J. Pediatr.* 1998, vol. 157, no. 7, p. 599-601.

ILLUMINATI, G., CALIÓ, FG., PAPASPYROPOULOS, V. et al. Revascularization of the internal carotid artery for isolated, stenotic, and symptomatic kinking. *Arch. Surg.* 2003, vol. 138, no. 2, p. 192-197.

JÄCKEL, M. Variations of the clinical course of the internal carotid artery – a differential diagnosis of parapharyngeal masses. *HNO.* 1997, vol. 45, no. 12, p. 1018-1021.

KELLY, AB. Tortuosity of the internal carotid in relation to the pharynx. *J. Laryngol. Otol.* 1925, vol. 40, p. 15-23.

LA BARBERA, G., LA MARCA, G., MARTINO, A. et al. Kinking, coiling, and tortuosity of extracranial internal carotid artery: is it the effect of a metaplasia? *Surg. Radiol. Anat.* 2006, vol. 28, no. 6, p. 573-580.

LE BRET, E., PINEAU, E., FOLLIGUET, T. et al. Congenital kinking of the internal carotid artery in twin brothers. *Circulation*. 2000, vol. 102, no. 22, p. 173-174.

METZ, H., MURRAY-LESLIE, RM., BANNISTER, RG. et al. Kinking of the internal carotid artery. *Lancet,* 1961, vol. 1, no. 7174, p. 424-426.

OKAMI, K., ONUKI, J., ISHIDA, K. et al. Tortuosity of the internal carotid artery – report of three cases and MR-angiography imaging. *Auris Nasus Larynx.* 2001, vol. 28, no. 4, p. 373-376.

OLIVIERO, U., COCOZZA, M., PICANO, T. et al. Prevalence of carotid kinking and coiling in a population at risk. *Vasc. Endovascular Surg.* 1997, vol. 31, p. 43-49.

ÖZBEK, C., YETKIN, U., ÖZELÇI, A. et al. Coiling of extracranial internal carotid artery as a cause of neurologic deficits. *The Internet Journal of Thoracic and Cardiovascular Surgery* 10, 2007, no.1.

PANCERA, P., RIBUL, M. and PRESCIUTTINI, B. Prevalence of carotid artery kinking in 590 consecutive subjects evaluated by echocolordoppler. Is there a correlation with arterial hypertension? *J. Intern. Med.* 2000, vol. 248, no. 1, p. 7-12.

PAULSEN, F., TILLMANN, B., CHRISTOFIDES, C. et al. Curving and looping of the internal carotid artery in relation to the pharynx: frequency, embryology and clinical implications. *J. Anat.* 2000, vol. 197, no. 3, p. 373-381.

PERDUE, GD., BARRECA, JP., SMITH, RB. et al. The significance of elongation and angulation of the carotid artery: a negative review. *Surgery.* 1975, vol. 77, no. 1, p. 45-52.

PRIOR, MJ., JOHNSON, IJM., JONES, K. et al. Aberrant common carotid artery encountered during free jejunal graft repair of esophageal stricture. *J Laryngol. Otol.* 1997, vol. 111, no. 8, p. 772-774.

QUATTLEBAUM Jr., JK., WADE, JS. and WHIDDON, CM. Stroke associated with elongation and kinking of the carotid artery: Long-term follow-up. *Am. Surg.* 1973, vol. 177, no. 5, p. 572-578.

RIBEIRO, RA., RIBEIRO, JAS., RODRIGUES FILHO, OA. et al. Common carotid artery bifurcation levels related to clinical relevant anatomical landmarks. *Int. J. Morphol.* 2006, vol. 24, p. 413-416.

SCHUMACHER, WA., SCHAFIG, A., KEHRL, W. et al. Variations of the course of the internal carotid artery – Potential risks in "standard operations" in the upper pharynx. *Laryngorhinootologie*. 1998, vol. 77, no. 9, p. 517-520.

SPADONI, E., NERI, F., LUTI, L. et al. Carotideal kinking and focal neurological symptoms. *Ital. J. Pediatr.* 2003, vol. 29, p. 309-311.

SZÉKELY, G. and CSÉCSEI, GI. Anteposition of the internal carotid artery for surgical treatment of kinking. *Surg. Neurol.* 2001, vol. 56, no. 2, p. 124-126.

TRACKLER, RT. and MIKULICICH, AG. Diminished cerebral perfusion resulting from kinking of the internal carotid artery. *J. Nucl. Med.* 1974, vol. 15, no. 7, p. 634-635.

WEIBEL, J. and FIELDS, WS. Tortuosity, coiling, and kinking of internal carotid artery. I Etiology and radiographic anatomy. *Neurology.* 1965, vol. 15, p. 7-18.

WITZ, M. and LEHMANN, JM. An association of internal carotid artery kinking and severe stenosis. *EJVES Extra.* 2001, vol. 1, p. 37-38.

Received May 7, 2008
Accepted October 6, 2008