Morphological Variation of the Carotid Arterial System Increases the Risk of Development of Central Nervous System Ischemia

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ABSTRACT: During a routine cervicothoracic dissection, an anomalous carotid vascular system was discovered. Anomalies of the carotid vascular system are clinically significant due to their principal role in supplying blood to the neck, head, and brain. Our findings reveal a severe morphological variation of the carotid vascular system, which significantly increases the risk of developing central nervous system ischemia. Variations in morphology, including kinking and tortuosity, of the carotid arterial system described in this study should be considered when evaluating the symptoms consistent with central nervous system ischemia. The individual studied suffered from dementia as well as a past medical history of cardiovascular accident (CVA), Hypertension (HTN), and depression, which can be clinically related to the morphological variations seen in the carotid arterial system.

KEYWORDS: carotid, carotid kinking and tortuosity, CNS ischemia, vascular anomalies, vertebral and carotid variations, dementia

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Introduction

Classic anatomy describes the aortic arch giving rise to the brachiocephalic trunk, left common carotid, and left subclavian artery, with the brachiocephalic trunk superiorly bifurcating into the right subclavian artery and the right common carotid artery. At approximately the level of the third cervical vertebra, the common carotid arteries bifurcate into the internal and external carotid. The external carotid primarily supplies the superficial portions of the head, scalp, face, and skull, while the internal carotid supplies the brain (Fig. 1). Embryological development can contribute to vascular anomalies, including tortuous kinks or coils, within individuals. Tortuosity is defined as any elongation, or S- or C-shaped curve of the vessel.[1] Kinking is described as angulation of one or more segments of an artery. Kinking of the carotid blood vessel system is commonly associated with stenosis in the affected segment.[1–3] Kinking has been graded as type I in the presence of an angle ranging from 60° to 90°, as type II in the presence of an angle ranging from 30° and 60°, and as type III (sharp) in the presence of an angle less than 30°.[1,4] Previous studies have shown that patients presenting with neurologic manifestations had prevalence rates of 54% and 44.6% for kinking and tortuosity, respectively, while coiling is more rare.[4] Kinking of the internal carotid artery is almost always accompanied by atherosclerosis with subintimal deposits, loss of elasticity, and elongation. The rate of aneurysmal development is also increased.[5] Coiling is commonly caused by elongation and redundancy of the carotid arterial system, resulting in an exaggerated S-shaped curvature or in a circular configuration.[1–3] It has been demonstrated that in human beings the internal carotid arteries carry approximately 90% of cerebral blood flow.[6] Cerebrovascular insufficiency associated with coils and kinks has been attributed to atherosclerosis due to abnormal embryologic development of the carotid arterial system.[5]

Case

During a routine cervicothoracic dissection of a 96-year-old female cadaver, an anomalous carotid vascular system was observed. Tortuosity of the bilateral common carotid arteries was noted secondary to elongation with S- and C-shaped curvatures. The brachiocephalic artery was somewhat tortuous and kinked upon itself (Fig. 2). Another significant finding involved severe kinking of the left common carotid artery with displacement medially, which caused the artery to become wedged between the esophagus and vertebral bodies of the mid to lower cervical spine (Fig. 3). After removing the carotid vascular system, the vertebral arteries were visualized. There was noted tortuosity and abnormal spinal level entrance

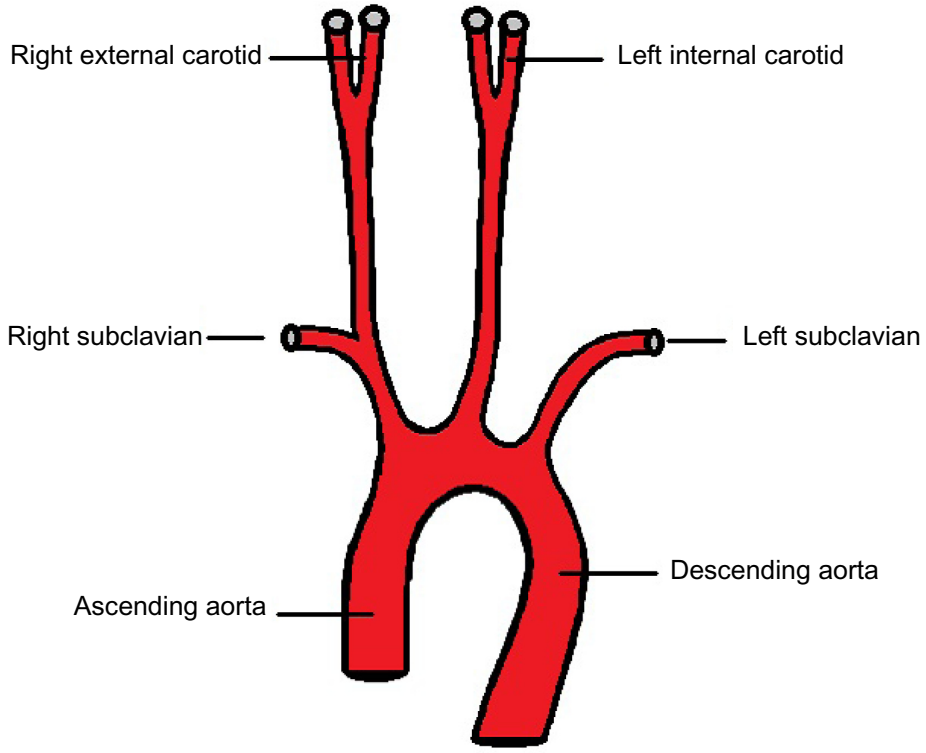

Figure 1. Aortic diagram. The aorta and associated branches of the carotid arterial system arising from the arch of the aorta.

into the associated intervertebral foramina. The left vertebral artery entered the intervertebral level between C5 and C6, while the right entered between C2 and C3 (Fig. 4). This was accompanied by severe S-shaped tortuosity at the level of intervertebral foramina entrance. Minor vascular dissimilarities in patients are common because the circulatory system will anatomically adapt as needed based on demand,[7] but the specific anomalous vascular dissimilarity we discovered is rarely observed during routine dissections.

Notably as it relates to our case, upon reviewing the certificate of death, this individual suffered from dementia and had a past medical history of CVA, HTN, and depression, which could be directly related to the condition outlined above.

Discussion

Embryogensis. During the middle of the third week, the vascular system of the embryo appears. In the fetus, one of the initial stages of vascular development involves the formation of aortic arches from the aortic sac during the fourth and fifth weeks. These aortic arches eventually go on to become many of the vascular structures in the neck, head, and brain. The aortic sac contributes a branch to each new arch with the formation of the successive pharyngeal arches. This development gives rise to a total of six pairs of arteries (Fig. 5A).

The ventral portion of the third aortic arch forms the common carotid and external carotid artery, while the dorsal portion forms the internal carotid artery. The cranial portion of the dorsal aorta forms the remainder of the internal carotid. The dorsal aorta connecting the third arch to more inferior structures begins to degenerate. Once this process is complete, the paired third arches along with the superior dorsal aorta become the right and left carotids (Fig. 5B), sending blood flow to the anterior and middle portions of the developing fetal brain.[8]

Throughout this entire process, the more rostral end of the dorsal aorta continues to develop and grow. As part of this process, they develop structures called intersegmental arteries that help vascularize the developing fetal spine. Over time, these intersegmental arteries elongate and join to begin sending blood flow to the posterior portion of the developing fetal brain. This vascular tissue eventually matures to become the vertebral arteries.[9]

Many dissimilarities of the carotid arterial system can be explained by derangements of embryological development,[10] associated with congenital anomalies of the aortic arch arteries.[11] Persistence and/or failure of aortic arch embryological absorption may lead to vessel elongation and subsequent curving, kinking, tortuosity, and looping, as presented in our study.[12] Additionally, multiple etiologies other than embryological development can cause kinking and coiling of the carotid arterial system, including age, arteriosclerosis, stenosis, vasculitis, and loss of elasticity.[13]

Clinical manifestations. Anomalies of the carotid arteries are clinically significant due to their principal role in supplying blood to the neck, head, and brain.[10] Variations of the carotid arteries, such as kinking, can result in

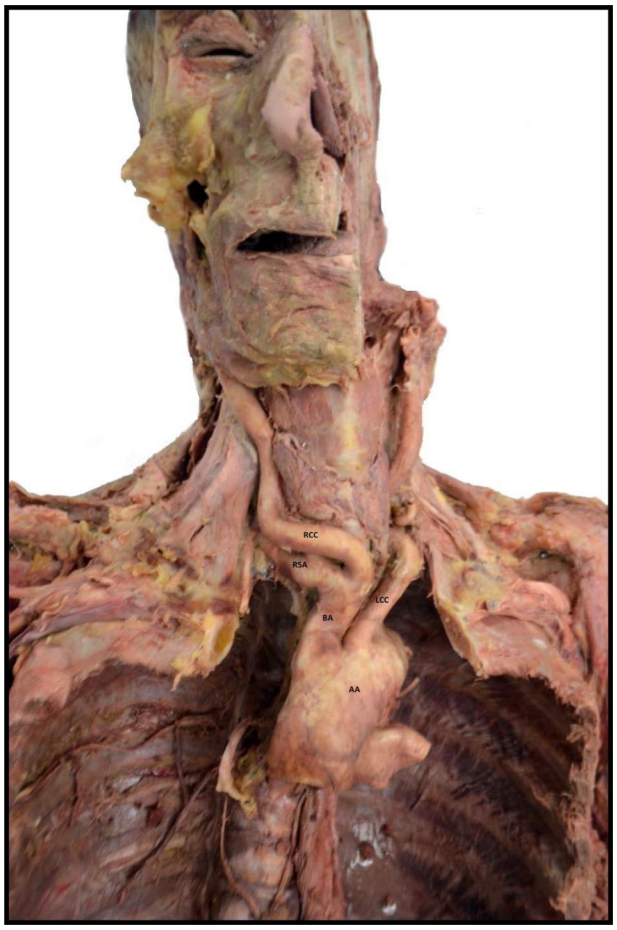

Figure 2. Tortuosity of the bilateral common carotid arteries is seen secondary to elongation with S- and C-shaped curvatures. The brachiocephalic artery is also visualized and kinked upon itself.
Abbreviations: RCC, right common carotid; LCC, left common carotid; RSA, right subclavian artery; BA, brachiocephalic artery; AA, aortic arch.

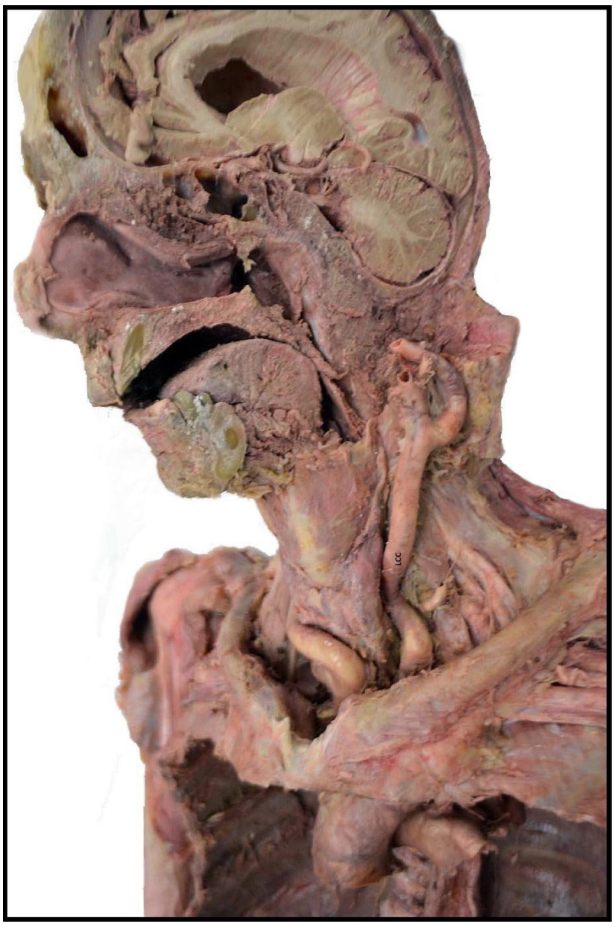

Figure 3. Note severe kinking of the LCC artery with displacement medially and wedged between the esophagus and vertebral bodies of the mid to lower cervical spine.
Abbreviation: LCC, left common carotid.

neurological symptoms. Symptoms reported include ischemic stroke or transient ischemic attack, carotid bruit, syncope, visual changes including amaurosis fugax, vertigo, mental status changes, or obscure neurological symptoms.[14] An abnormal curvature may produce luminal narrowing or lead to turbulent blood flow, resulting in secondary ulceration of the intima or formation of an embolus.[15] Concomitant disease states include atherosclerotic disease, hypertension, coronary artery disease, and peripheral artery disease.[10,14,16] Anomalies of this type may present as a pulsatile mass, which could mimic an aneurysm.[10] Anomalies should be considered in patients who report neurological symptoms without an identifiable cause and in young, or otherwise healthy, patients. Symptoms are likely exacerbated during ipsilateral rotation, extension, and/or contralateral cervical spine lateral bending. Reduced blood flow with insubstantial compensation from the vertebral arteries is the suspected cause of increased symptomatology.[15] While the authors believe that the kinking of the carotid arteries may cause or contribute to neurologic symptoms, it is important to entertain a broad spectrum of differential diagnoses. This manifestation can be best explained by Metz et al.

It is always difficult to be certain of the relevance of any factor to the production of cerebrovascular symptoms.

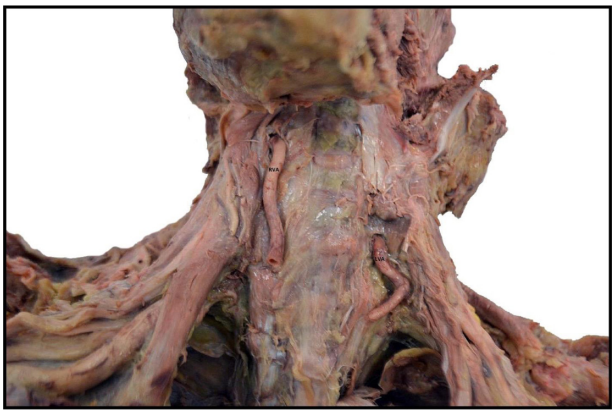

Figure 4. Vertebral arteries visualized with noted tortuosity and abnormal spinal level entrance. Left vertebral artery enters the intervertebral level between C5 and C6, while the right entered between C2 and C3.
Abbreviations: RVA, right vertebral artery; LVA, left vertebral artery.

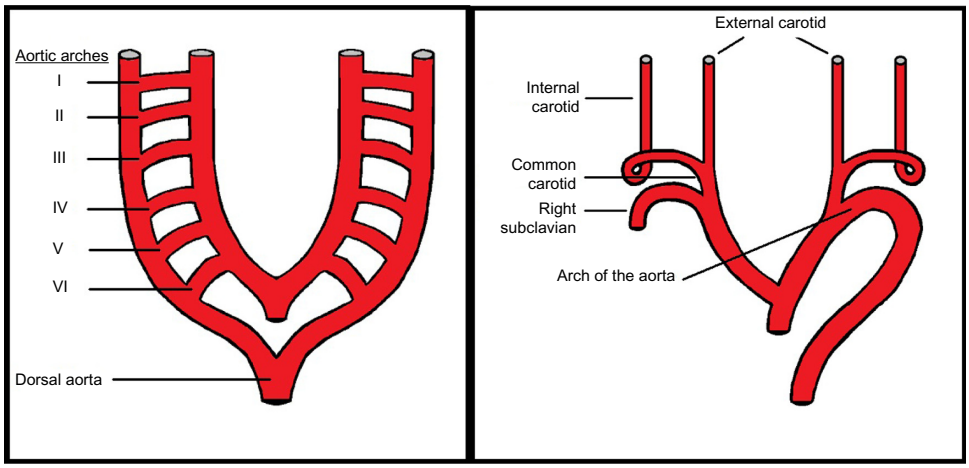

Figure 5. Aortic arches and dorsal aorta before (**A**) and after (**B**) transformation into definitive vascular pattern.

Cessation of symptoms after removal of a factor does not establish the existence of a causal relation between the two. It is well recognized, for example, that transient ischemic episodes may cease spontaneously; hence, the disappearance or nonrecurrence of symptoms following the excision of a kink in the internal carotid artery must be interpreted with caution. Nevertheless, the present unsatisfactory state of therapy for cerebrovascular disease demands that each potential method of treatment be thoroughly assessed.[4]

Kinking of the carotids may explain neurologic symptoms; however, this anatomical variation is like any other disease or disorder in medicine: not all patients will present the same way. A patient with kinked carotids could present asymptomatically and it would be found incidentally, or not at all.

Carotid abnormalities become especially significant when performing surgeries of the neck. A midline vertical neck incision for a tracheostomy could cause increased bleeding if the common carotid artery crosses the trachea and is injured.[10] Other invasive procedures could result in an encounter of an aberrant carotid artery, as well. In addition, when the common carotid artery bifurcates more superiorly, as in the case mentioned, the common carotid artery is located closer to the hypoglossal nerve, which puts the nerve at an increased risk of damage.

Diagnosis and treatment. An anomalous carotid artery can be diagnosed via several different methods. Angiography is considered the gold standard.[16,17] Ultrasound, magnetic resonance angiogram, computed tomography with contrast, and magnetic resonance imaging have also been used to diagnose variations in the carotid arterial system.[1,14,18–20] An electroencephalogram can also be valuable if symptoms are exacerbated during head rotation.[16]

Because of the neurologic symptoms that may result from a kinked carotid artery, surgery is sometimes necessary to correct the abnormality. Surgical intervention should be selective with increased indication in patients with paroxysmal neurological symptoms, especially during rotation of head.[16] Head movements should be performed during arteriography to visualize reduced areas of blood flow caused by the kinking.[5]

Vannix et al suggested the following criteria for surgery of the carotid arteries to relieve neurologic symptoms[5]:

- The patient should show definite central nervous system symptoms without other arteriographic abnormalities.
- The lumen is at least 40%–50% of diameter of the blood vessel.
- Movement or acute rotation of the head reproduces and/or exaggerates symptoms.
- A brain scan does not reveal abnormal pathology.
- No evidence of atherosclerotic disease is present at the bifurcation of the common carotid artery.

The appropriate surgery would be an end-to-end anastomosis after resection of common carotid artery to correct the kink or break adhesions that produce or contribute to the kink.[16] Patients with neurologic symptoms, found to be caused by kinks in the carotid arterial system, who underwent surgery generally had positive outcomes.[15–17,20]

Conclusion

Variation of the carotid arterial system more frequently involves the internal carotid artery[1,18,19] with the right common carotid found to be more tortuous than the left.[10,21] However, the case we present had a severe, multidirectional type III kinking of the left common carotid artery.

Anomalous variations such as elongations, tortuosity, coiling, kinking, or looping are commonly a consequence of deranged embryological development. Patients with neurologic manifestations were found to have a higher prevalence of kinking than tortuosity or coiling.[4] Kinking increases the risk of developing arteriosclerosis, stenosis, vasculitis, atrophic dilation, loss of elasticity, and dissection in the context of fibromuscular dysplasia.[13] These types of variations often

go undetected in the affected populations and are benign unless symptoms develop. It has been found that patients with neurologic manifestations, resulting from carotid arterial system abnormalities, benefit greatly from surgery.[15–17,20] It is the opinion of the authors that variations in morphology of the carotid arterial system should be considered when evaluating symptoms consistent with central nervous system ischemia.

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Author Contributions

Conceived and designed the experiments: EC, SBM, NP, LK, MJC. Analyzed the data: EC, SBM, NP, MJC. Wrote the first draft of the manuscript: EC, SBM, MJC. Contributed to the writing of the manuscript: EC, SBM, NP, LK, MJC. Agreed with manuscript results and conclusions: EC, SBM, NP, LK, MJC. Jointly developed the structure and arguments for the paper: EC, SBM, NP, LK, MJC. Made critical revisions and approved the final version: EC, SBM, NP, LK, MJC. All the authors reviewed and approved the final manuscript.

REFERENCES

1. Sacco S, Totaro R, Baldassarre M, Carolei A. Morphological variations of the internal carotid artery: prevalence, characteristics and association with cerebrovascular disease. *Int J Angiol*. 2007;16(2):59–61.
2. Weibel J, Fields WS. Tortuosity, coiling and kinking of the internal carotid artery I: etiology and radiographic anatomy. *Neurology*. 1965;15:7–18.
3. Weibel J, Fields WS. Tortuosity, coiling and kinking of the internal carotid artery II: relationship of morphological variation to cerebrovascular insufficiency. *Neurology*. 1965;15:462–8.
4. Metz H, Bannister RG, Murray-Leslie RM, Bannister RG, Bull JW, Marshall J. Kinking of the internal carotid artery. *Lancet*. 1961;1:424–6.
5. Vannix RS, Joergenson EJ, Carter R. Kinking of the internal carotid artery: clinical significance and surgical management. *Am J Surg*. 1977;134:82–9.
6. Roberts B, Hardesty WH, Holling HE, Reivich M, Toole JF. Studies on extracranial cerebral blood flow. *Surgery*. 1964;56:826–33.
7. Ciani M, Gonzalez M, Henson T, Young K, Ciani G. Considering an aortic arch anomaly in patients with dysphagia. *JAAPA*. 2013;26(11):53–7. doi: 10.1097/01.JAA.0000436521.53922.7c.
8. Sadler TW. Cardiovascular system. *Langman's Medical Embryology*. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2012:185–90.
9. Satti SR, Cerniglia CA, Koenigsberg RA. Cervical vertebral artery variations: an anatomic study. *AJNR Am J Neuroradiol*. 2007;28:976–80.
10. Shankar N, Raveendranath V, Ravindranath R, Manjunath KY. Anatomical variations associated with the carotid arterial system in the neck. *Euro J Anat*. 2008;12:175–8.
11. Parkinson J, Bedford DE, Almond S. The kinked carotid artery that simulates aneurysm. *Br Heart J*. 1939;1:345–61.
12. Ovchinnikov NA, Rao RT, Rao SR. Unilateral congenital elongation of the cervical part of the internal carotid artery with kinking and looping: two case reports and review of literature. *Head Face Med*. 2007;3:29.
13. Schenk P, Temmel A, Trattnig S, Kainberger F. Current aspects of diagnosis and therapy of carotid artery kinking. *HNO*. 1996;44(4):178–85.
14. Togay-Isikay C, Kim J, Betterman K, et al. Carotid artery tortuosity, kinking, coiling: stroke risk factor, marker, or curiosity? *Acta Neurol Belg*. 2005;105(2):68–72.
15. Milic DJ, Jovanovic MM, Zivic SS, Jankovic RJ. Coiling of the left common carotid artery as a cause of transient ischemic attacks. *J Vasc Surg*. 2007;45(2):411–3.
16. Leipzig TJ, Dohrmann GJ. The tortuous or kinked carotid artery: pathogenesis and clinical considerations. A historical review. *Surg Neurol*. 1986;25:478–86.
17. Desai B, Toole JF. Kinks, coils, and carotids: a review. *Stroke*. 1975;6:649–53.
18. Muñoz A, De Vergas J, Crespo J. Imaging and clinical findings in patients with aberrant course of the cervical internal carotid arteries. *Open Neuroimag J*. 2010;4:174–81. doi: 10.2174/1874440001004010174.
19. Tse GG, Masuda EM, McMurtray AM, Nakamoto BK. Coiled internal carotid arteries associated with bilateral sequential strokes. *Case Rep Vasc Med*. 2013;2013:929530. doi: 10.1155/2013/929530.
20. Tetik O, Yurekli I, Yilik L, Akhan G, Gurbuz A. Surgical treatment of symptomatic coiling or kinking internal carotid artery. *Vascular*. 2010;18(5):294–6.
21. Fazan VP, Ribeiro RA, Oliveira MA, Caetano AG, Rodrigues Filho OA. Tortuosity of the internal carotid artery cervical course: case reports and literature review. *Braz J Morphol Sci*. 2007;24(4):244–7.