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Nonbifurcating Carotid Artery: A Case Report with a Review of Embryogenesis

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 66
Final Diagnosis: Nonbifurcating carotid artery
Symptoms: No symptoms
Medication: —
Clinical Procedure: —
Specialty: Neurology

Objective: Congenital defects/diseases

Background: Vascular anomalies of the carotid vessels can be attributed to false embryogenesis. A rare variant called a nonbifurcating carotid artery (NBCA) exists, where typical carotid bifurcation is not recognizable with its typical branches of the external carotid artery (ECA) and internal carotid artery (ICA). This paper describes a case of this anomaly and reviews the embryogenesis of the carotid arteries for explanation.

Case Report: A 66-year-old man received a routine health examination at our hospital. Initial carotid ultrasound indicated an absence of bifurcation in the right cervical carotid artery, and magnetic resonance imaging of the brain indicated an absence of the proximal cervical segment of the right ICA, with a remnant arterial stump at the expected bifurcation level. No evidence of the carotid bulb was identified. The common carotid artery seemed to continue cranially in the trunk of the ECA, where it exhibited extracranial branches. After distributing these branches, the carotid artery coursed medially at the C2 level, where it ascended into the carotid canal to become the petrosal segment of the ICA. This carotid anomaly was labelled an NBCA. No aberrant intracranial arteries were derived from the NBCA in this case.

Conclusions: In this case, the arterial stump was considered a remnant from agenesis of the right ICA. We assumed that the NBCA most likely developed because of false regression of the third embryogenic aortic arch with persistence of the second aortic arch.

MeSH Keywords: Cardiovascular System • Carotid Arteries • Embryology

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Background

Carotid ultrasound and magnetic resonance imaging (MRI) are 2 commonly used noninvasive techniques for evaluating the carotid vessels. With the growing popularity of these tools in general health examinations, increasingly more anomalies of the common carotid artery (CCA) are being discovered. Among them, a rare anomaly exists in which the carotid bifurcation is indistinguishable from its typical branches of the external carotid artery (ECA) and internal carotid artery (ICA). This anomaly is referred to as a nonbifurcating carotid artery (NBCA) and was first proposed by Marimoto et al. in 1990 [1]. In a retrospective cohort study conducted in Japan, the estimated incidence of NBCAs was 0.21%, which was almost identical to the reported 0.2% incidence of persistent trigeminal arteries [2]. In an NBCA, the carotid bulb is absent in the cervical segment; this is suggestive of proximal ICA agenesis. This paper describes a case of an NBCA and explains its possible embryogenesis.

Case Report

A 66-year-old man with hypertension and dyslipidemia under medical control underwent imaging studies during a health examination at our hospital. A carotid ultrasound indicated an abrupt blockade of the right proximal ICA with a small residual arterial stump at the bifurcation. Neither atherosclerotic plaque nor turbulent flow was discovered in the vascular lumen (Figure 1). Therefore, the patient underwent brain MRI for the suspicious total occlusion of the right ICA. Magnetic resonance angiography (MRA) revealed that the right CCA was considerably enlarged at the C4 to C5 level to a size equivalent to that of the left carotid bulb, but did not taper 1–2 cm distally, as a healthy carotid bulb would. Moreover, no typical carotid bifurcation was present – only an arterial stump originating from the medial aspect of the right CCA (Figure 2). The right CCA continued in the trunk with 3 major extracranial branches: the common trunk of the superior thyroid artery and facial artery, the occipital artery, and the common trunk of the internal maxillary artery and posterior auricular artery. After distribution from the extracranial branches, the right CCA coursed medially at the C2 level to enter the right carotid canal, where it became the petrosal segment of the right ICA. The intracranial course of the right ICA was typical, and the left ECA, ICA, and vertebral artery (VA) were typical in appearance. The right VA was atrophic and the left VA was dominant in the posterior cerebral circulation. Diffusion-weighted imaging and brain perfusion imaging produced unremarkable findings.

Discussion

The embryogenesis of vascular systems in the head and neck is complicated. Early in the third gestational week, the cervical

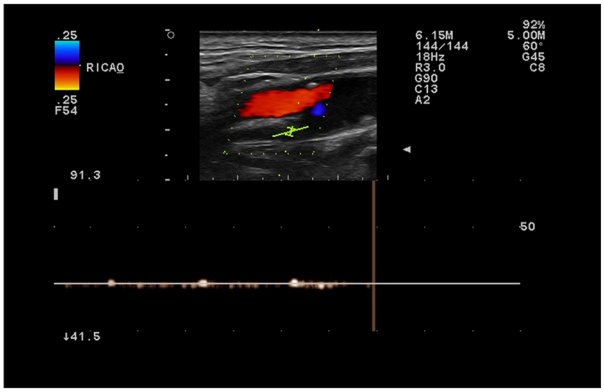

Figure 1. Carotid color Doppler sonography of the NBCA. The right proximal ICA is abruptly terminated without vascular flow in the lumen. Notably, an arterial stump is present at the expected bifurcation level.

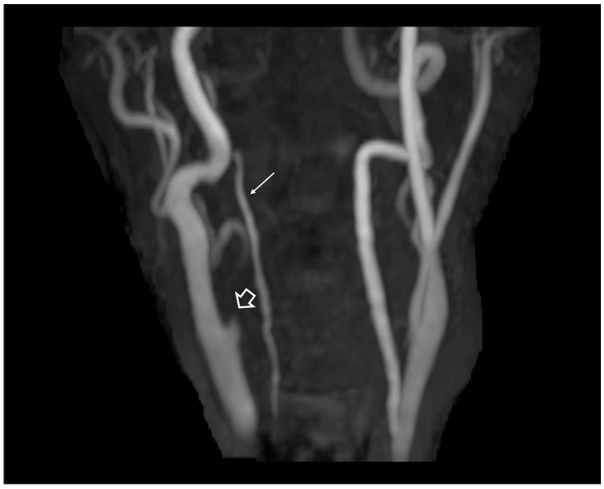

Figure 2. Three-dimensional reconstruction of the NBCA on time-of-flight MRA. Loss of bifurcation in the right carotid artery with an arterial stump located at the expected bifurcation (open arrow). The CCA seems to continue in the trunk of the ECA and gives off 3 major extracranial branches. Hypoplasia of the right VA is also demonstrated in the image (arrow).

aortic primitives run in pairs as dorsal and ventral aortic roots. Six primitive aortic arches connect the dorsal and ventral aortic roots, and 6 intersegmental arteries run laterally from the dorsal aortic roots. The ventral aortic roots cranial to the third aortic arch become the ventral pharyngeal artery. The ventral second aortic arch eventually regresses. The dorsal second aortic arch becomes the hyoid artery and stapedial artery. The stapedial artery connects the ventral pharyngeal artery and separates from the hyoid artery at the 24-mm embryonic stage (approximately the seventh gestational week). The ECA develops from the combination of the ventral pharyngeal artery and stapedial artery. The ICA derives from the third aortic arch and the dorsal aortic roots cranial to the third aortic arch.

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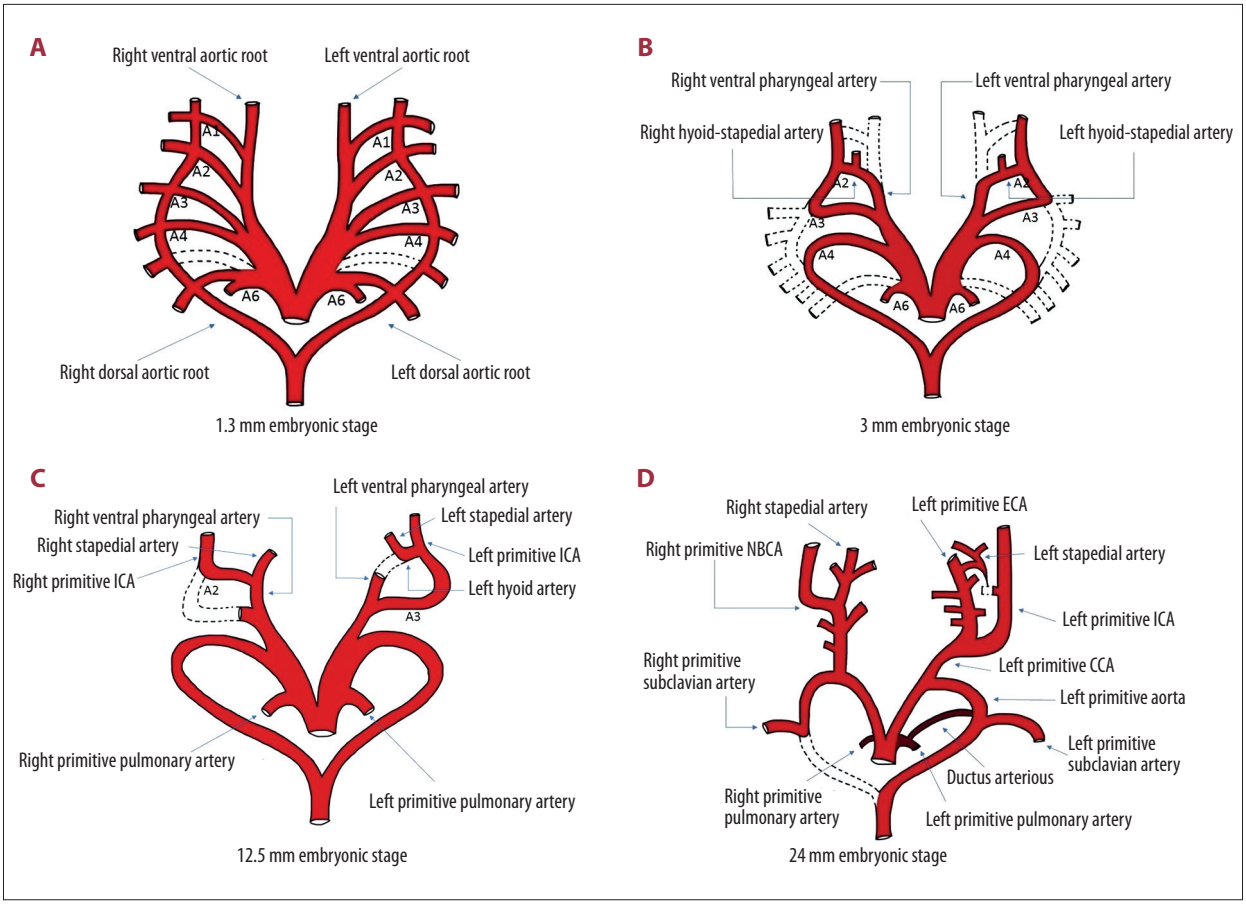

Figure 3. Speculated embryogenesis of the NBCA. (**A**) At the 1.3-mm embryonic stage (third gestational week), the dorsal and ventral aortic roots run in pairs with 6 aortic arches crossing over them. Six intersegmental arteries run laterally from the dorsal aortic roots. (**B**) At the 3-mm embryonic stage, the ventral aortic roots cranial to the third aortic arch become the ventral pharyngeal artery. The dorsal second aortic arch becomes the hyoid–stapedial artery. (**C**) At the 12.5-mm embryonic stage, the left ventral second aortic arch regresses but the right ventral second aortic arch persists as a connection between the primitive ECA and ICA. The right third aortic arch regresses at this stage. (**D**) The left ECA and ICA develop typically at the 24-mm embryonic stage (seventh gestational week). The right NBCA develops from the linkage of the ventral pharyngeal artery and the second aortic arch.

The CCA evolves from the fusion of the ventral pharyngeal artery and ventral third aortic arch. The VA develops from the transverse anastomoses between the cervical intersegmental arteries [3]. In our case, the embryogenesis of the left carotid artery followed the typical development process (Figure 3).

We hypothesized that the right NBCA coursed in the trunk of the proximal ECA and then turned medially at the C2 level to become the petrosal segment of the ICA. Our imaging findings supported this hypothesis. First, the carotid bulb was absent in the right NBCA, and thus it was unlikely that the right CCA coursed cranially in the trunk of the ICA, which usually forms a bulb in the proximal segment. Second, almost all extracranial branches, which are the branches of the ECA under typical conditions, could be identified in the NBCA in our case. Third, the NBCA coursed with a great tortuosity at the C2 level; this evidenced the anastomosis between the ECA and ICA. Therefore, we ascertained that the NBCA coursed cranially in the trunk of the ECA with an anastomosis with the ICA at the C2 level. Moreover, 3 extracranial branches derived from the stapedial artery were identifiable: the distal internal maxillary artery, the posterior auricular artery, and the occipital artery. Because the stapedial artery originated from the dorsal second aortic arch, we deduced that the second aortic arch persisted as a connection between the ventral and dorsal aortic roots. The dorsal third aortic arch regressed, thereby causing the ventral third aortic arch to become an arterial stump in the NBCA (Figure 3).

Our case shared 2 common features identified in most published studies regarding agenesis of the proximal ICA. First, nearly all extracranial branches of the NBCA indicate identical courses to the healthy branches derived from the ventral pharyngeal artery and stapedial artery, which are the primitives of

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Table 1. Imaging findings of nonbifurcating carotid arteries in relevant case reports.

Authors (year)	Extracranial branches derived from the stapedial artery	Carotid bulb	Aortic stump	Intracranial ICA anomalies
Morimoto et al. (1990)	IMA, OA	(–)	(–)	(–)
Lambiase et al. (1991)	IMA, SFTA	(–)	(–)	(–)
Katsuji et al. (1996)	IMA, PAA, OA	(–)	(–)	N/A
Ooigawa et al. (2006)	IMA, OA	(–)	(–)	(–)
	OA	(–)	(–)	(–)
Kiyosue et al. (2009)	IMA, PAA, OA	(–)	(–)	Dural arteriovenous fistula
Iimura et al. (2009)	IMA, OA	(–)	(–)	Anomalous connection between CCA and ICA
Murono et al. (2009)	SFTA, IMA	(–)	(–)	(–)
Uchino et al. (2011)	OA	(–)	(–)	(–)
Sasaki et al. (2013)	MMA	(–)	(+)	(–)
Lourenco et al. (2014)	IMA, PAA, OA	(–)	(–)	(–)
Kim et al. (2015)	IMA, OA	(–)	(+)	(–)
Yoshida et al. (2016)	IMA, OA	(–)	(–)	(–)

IMA – internal maxillary artery; PAA – posterior auricular artery; OA – occipital artery; MMA – middle meningeal artery; SFTA – superficial temporal artery; CCA – common carotid artery; ICA – internal carotid artery.

ECA [1,2,4,5]. Second, the proximal course of most NBCAs resembles that of the ECA, and the tortuosity at the C1 to C2 level suggests a connection with the primitive ECA and ICA [2,6]. These features support the hypothesis that an NBCA develops from agenesis of the proximal ICA, with an anastomosis of the distal ECA and ICA. In addition, the intracranial course of most NBCAs resembles that of a typical distal ICA, which is developed from segments distal to the second dorsal aortic root [3,6–8]. Thus, we considered the possibility that the NBCA develops typical aortic roots distal to the second aortic arch. Some cases have described NBCAs without aortic stumps (Table 1) [4,8,9]. Therefore, we suspected that the starting point of the regression in the third aortic arch might have been different in these cases.

Regarding related genetic factors, 2 homeobox genes, Hoxa-2 and Hoxa-3, have been reported to influence the development of the second and third aortic arches in animal studies [10–13]. Osamu et al. discovered a highly specific defect of the third pharyngeal arch development in Hoxa-3 null mutant mice. In their experiment, the initial formation of the third arch artery was not disturbed but the artery regressed at embryonic day 11.5 when differentiation of the third pharyngeal arch began. In the mice, the ECA and ICA originated from the fourth aortic root and an NBCA was not developed [11]. However, it was proven that overexpression of Hoxa-2 resulted in a transformation of first arch structures into second arch elements [13]. Nevertheless, there is no direct evidence that persistence of the second aortic arch is related to Hoxa-2 overexpression. Therefore, the exact genetic disorders related to NBCAs remain unknown.

Carotid atherosclerosis can coexist with NBCAs [4,9]. If the patient also presents severe atherosclerotic stenosis, treatment with carotid artery stenting or carotid endarterectomy is favorable; both treatments require an adequate distal protection device to reduce the risk of thromboembolic events [9,14]. Recurrent cerebrovascular events can originate from an arterial stump in a similar condition called carotid stump syndrome [15]; therefore, the risk of thromboembolic events during procedures cannot be overlooked in cases of an NBCA combined with an arterial stump, because the arterial stump may produce a turbulent flow in the NBCA. For patients in need of carotid intervention, preoperative evaluation of the branching pattern and stenosis region helps the surgeon in selecting a suitable distal protection device [14]. The surgeon should be familiar with collateral circulation and regional hemodynamics when stenting or endarterectomy is performed on an atherosclerosis-related stenotic NBCA.

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Conclusions

In our case, the right NBCA was formed due to ICA agenesis and an anastomosis was present between the right distal ECA and cervical segment of the right ICA. A congenital anomaly such as this is most likely to develop because of false regression of the third embryogenic aortic arch with persistence from the second aortic arch. Which mechanism leads to such an unusual development of the carotid artery remains unclear. In clinical practice, perioperative thromboembolic events should be considered when stenting or endarterectomy is performed in an atherosclerosis-related stenotic NBCA.

Acknowledgements

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Conflicts of interest

The authors declare no conflicts of interest.

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