Carotid Artery Stenting for Atherosclerotic Stenosis Associated With Non-bifurcating Cervical Carotid Artery
—Case Report and Embryological Considerations—

Tetsuo SASAKI,1 Hisashi NAGASHIMA,2 Fusakazu OYA,3 Daisuke SATOH,1 and Shigeaki KOBAYASHI4

1Interventional Neuroradiology Center, and
4Department of Neurosurgery, Aizawa Hospital, Matsumoto, Nagano;
2Interventional Neuroradiology Center, Shinshu University Hospital, Matsumoto, Nagano;
3Department of Neurosurgery, Nagano Municipal Hospital, Nagano, Nagano

Abstract
A 68-year-old man presented with right cervical carotid artery stenosis manifesting as ipsilateral amaurosis fugax. Angiography showed non-bifurcating cervical carotid artery with atherosclerotic stenosis near the branching of the superior thyroid artery. Carotid artery stenting (CAS) was carried out using two balloons for embolic protection of the internal carotid artery and middle meningeal artery because the ophthalmic artery was supplied by the middle meningeal artery. No procedure-related complications or restenosis occurred after stenting. Non-bifurcating cervical carotid artery is a very rare anomaly, in which the branches of the external carotid artery directly arise from the common trunk of the carotid artery without forming a bifurcation. The present patient was successfully treated with CAS for atherosclerotic stenosis associated with non-bifurcating cervical carotid artery.

Key words: anomaly, carotid artery stenting, carotid bifurcation, non-bifurcating cervical carotid artery, segmental agenesis

Introduction
Non-bifurcating cervical carotid artery is a very rare anomaly of the cervical carotid artery, in which the branches of the external carotid artery (ECA) directly arise from the common trunk of the carotid artery without forming a bifurcation.7,14,15,19,22 We describe here the case of a patient successfully treated with carotid artery stenting (CAS) for atherosclerotic stenosis associated with non-bifurcating cervical carotid artery and discuss the presumed embryological development of this anomaly.

Case Report
A 68-year-old man presented with amaurosis fugax in the right eye and visited a local outpatient clinic. His medical history included heavy smoking, and hypertension and angina pectoris treated medically. Duplex ultrasonography revealed hypoechoic plaque in the right common carotid artery (CCA), and the patient was referred to our department. Fundus biomicroscopy revealed no abnormal findings. Magnetic resonance (MR) imaging showed asymptomatic ischemic lesions in the right frontal and temporal lobes. Right common carotid arteriography showed atherosclerotic stenosis of 55% near the branching of the superior thyroid artery (Fig. 1A, B). Angiography also revealed absence of the right carotid bifurcation, all branches of the ECA arose from the common trunk of the carotid artery, and continuation of the common trunk as the internal carotid artery (ICA) that entered the cranial cavity as usual.

CAS was carried out for the stenotic lesion coexisting with this anomaly, non-bifurcating cervical carotid artery, using two balloons for embolic protection of the ICA and middle meningeal artery because the right ophthalmic artery was supplied by the middle meningeal artery (Fig. 1C). A 7 French ultralong sheath (Shuttle; Cook, Bloomington, Indiana, USA) was advanced to the right CCA. To prevent embolic events in the ophthalmic artery and ICA territory, a PercuSurge GuardWire (Medtronic, Santa Rosa, California, USA) was placed in the ICA, and a 3.5 × 10 mm balloon (Sentry; Boston Scientific, Natick, Massachusetts, USA) was placed in the internal maxillary artery branching from the middle meningeal artery because the right ophthalmic artery was supplied by the middle meningeal artery (Fig. 1D). After predilating the lesion with a 5.0 × 20 mm balloon (Gazelle; Boston Scientific) at 8 atm, a 8 × 40 mm nitinol self-expandable stent (SMART; Cordis, Miami Lakes, Florida, USA) was placed across the lesion and deployed. Postdilation was not performed because the stent was successfully deployed without residual stenosis (Fig. 1E). Postoperative course was uneventful. No restenosis of the lesion, stroke, or amaurosis fugax was ob-
Fig. 1 Digital subtraction angiograms, anteroposterior view (A) and lateral views (B–E), of the present case. A, B: Right common carotid arteriograms showing non-bifurcating cervical carotid artery and atherosclerotic stenosis near the branching of the superior thyroid artery (at the level of C3-4). C: Ophthalmic artery was supplied by the middle meningeal artery (arrowheads). D: Carotid artery stenting was carried out using two balloons (arrows) for protection of the internal carotid and middle meningeal arteries because the right ophthalmic artery was supplied by the middle meningeal artery. E: Stent was successfully deployed without residual stenosis.

Fig. 2 A–C: Schematic drawings of the development of the cervical carotid artery. The second aortic arch regresses and becomes the hyoid artery. The stapedial artery, the main branch of the hyoid artery, passes through the middle ear and forms the middle meningeal and internal maxillary arteries. The ventral aorta becomes the ventral pharyngeal artery, which forms the facial and lingual arteries and connects to the internal maxillary artery. The internal carotid artery (ICA) originates from the third aortic arch and dorsal aorta. The ICA is constituted of 7 segments according to the “segmental identity” proposed by Lasjaunias. D: Non-bifurcating cervical carotid artery is probably formed by agenesis of the proximal part of the cervical ICA. A-I: first aortic arch, A-II: second aortic arch, A-III: third aortic arch, DA: dorsal aorta, DC: ductus caroticus, DO: dorsal ophthalmic artery, FL: facial and lingual arteries, hy: hyoid artery, IM: internal maxillary artery, m: mandibular artery, MM: middle meningeal artery, PM-PT: primitive maxillary and trigeminal arteries, st: stapedial artery, VA: ventral aorta, VO: ventral ophthalmic artery, Vph: ventral pharyngeal artery.

Discussion

Multiple anomalies of the carotid artery are known, but non-bifurcating cervical carotid artery is rarely described, with only 5 cases of non-bifurcating cervical carotid artery,7,14,15,19,22] and 6 similar cases with various terminologies.3,6,8,16,17,20] Two hypotheses have been described for the development of these anomalies, agenesis of the ECA trunk,5,8] and segmental agenesis of the ICA.7,14,17,19]

Non-bifurcating cervical carotid artery might be caused by absence of regression of the proximal hyoid artery with failure of transfer of its distal branches.8] Embryologically, the cervical carotid artery develops from the ventral aorta, dorsal aorta, and aortic arches (Fig. 2A). The second aortic arch regresses and becomes the hyoid artery. The stapedial artery, the main branch of the hyoid artery, passes through the middle ear and becomes the middle meningeal and internal maxillary arteries. The ventral aorta becomes the ventral pharyngeal artery, which forms the facial and lingual arteries and connects to the internal maxillary artery (Fig. 2B, C). If non-bifurcating cervical carotid artery is caused by agenesis of the ECA trunk with regression failure of the hyoid artery, the maxillary artery system should arise from the petrous portion of the ICA. In the present case, this hypothesis was not applicable because the internal maxillary artery arose from the cervical portion.

The ICA originates from the third aortic arch and dorsal aorta (Fig. 2A–C). According to the “segmental identity” proposed by Lasjaunias,9] the ICA is constituted of 7 seg-
ments (Fig. 2C). Each of these segments is located between embryonic arteries or their remnants, and can be absent, thus representing a focal agenesis. Therefore, non-bifurcating cervical carotid artery is probably expressed as a type of segmental agenesis of the ICA, especially agenesis of the cervical segment (Fig. 2D). In some cases,17 an arterial stump was observed as in our other case (Fig. 3), which would support this hypothesis.

Non-bifurcating cervical carotid artery may also be caused by maldevelopment of the third aortic arch with persistence of the first or second aortic arch,17 whereas the ICA constructed via the second aortic arch is known as aberrant ICA.2,11 It is generally thought that aberrant ICA is formed from the anastomosis between the tympanic branch of the ascending pharyngeal artery and carotico-tympanic artery, which is a dorsal remnant of the hyoid artery. The intracranial ICA is supplied by the cervical carotid artery through its anastomosis in the middle ear. However, the present case showed no aberrant course of the carotid artery.

The vessel separating between segments 1 and 2 is the ductus caroticus, which connects the third and fourth aortic arches. Persistent ductus caroticus with regression of the third aortic arch is thought to become another anomaly called absence of the CCA.13 Therefore, non-bifurcating cervical carotid artery caused by agenesis of the cervical ICA (segment 1) should use another embryonic artery as a collateral pathway excluding the ductus caroticus.

The occipital and ascending pharyngeal arteries develop from the proatlantal and hypoglossal arteries, respectively. According to Lasjaunias et al.,10,12 the occipital and ascending pharyngeal arteries arise from the primitive ICA and ECA, and the vertebral artery at points determined by the sites of regression. It is well known that the occipital artery can arise from the ICA, and the ascending pharyngeal artery arose from the ICA in a few cases.1,4 Consequently, segment 1 could further be divided into 2 or 3 parts by the branching points of the proatlantal or hypoglossal artery. Therefore, speculation would suggest that non-bifurcating cervical carotid artery is formed by agenesis of the proximal part of segment 1 using the pharyngo-occipital artery system as a collateral pathway (Table 1, Fig. 2D). Furthermore, we speculate that absence of the CCA and aberrant ICA result from agenesis of segment 1, and segments 1 and 2, respectively (Table 1).

Several cases of significant carotid artery stenosis associated with non-bifurcating cervical carotid artery have been reported.3,8,20 These cases were treated with carotid endarterectomy (CEA), but no cases were treated with CAS, as performed in the present patient with non-bifurcating cervical carotid artery (Table 2).

Cervical carotid artery stenosis is usually observed on the outer wall of the carotid sinus at the carotid bifurcation. Low wall shear stress due to hemodynamic conditions around the bifurcation is speculated to be one of the predisposing factors to induce atherosclerosis.18

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age (yrs), Sex</th>
<th>Presence of stroke</th>
<th>Plaque characteristics</th>
<th>Location of stenosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Franklin et al. (1988)13</td>
<td>43, M</td>
<td>asymptomatic</td>
<td>not evaluated</td>
<td>proximal to FLA and IMA (C2–C4)</td>
<td>CEA (stopped)</td>
</tr>
<tr>
<td>Lambiase et al. (1991)18</td>
<td>76, M</td>
<td>episodic confusion</td>
<td>typical atheroma without hemorrhage (specimen)</td>
<td>proximal to FLA and IMA (C3–4)</td>
<td>CEA</td>
</tr>
<tr>
<td>Rodriguez et al. (2002)20</td>
<td>66, M</td>
<td>asymptomatic</td>
<td>complex plaque (US)</td>
<td>proximal to FLA and IMA (C4)</td>
<td>CEA</td>
</tr>
<tr>
<td>Present case</td>
<td>68, M</td>
<td>amaurosis fugax</td>
<td>hypoechoic plaque (US)</td>
<td>proximal to FLA and IMA (C3–4)</td>
<td>CAS</td>
</tr>
</tbody>
</table>


Table 1 Congenital anomalies of the internal carotid artery (ICA)

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Proximal part of segment 1</th>
<th>Segments 1 and 2</th>
</tr>
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<tbody>
<tr>
<td>Collateral</td>
<td>proatlantal (OA) or hypoglossal (APA)</td>
<td>ductus caroticus</td>
</tr>
<tr>
<td>Anomaly</td>
<td>non-bifurcating cervical carotid artery</td>
<td>hypoglossal (APA)-hyoid (carotico-tympanic artery)</td>
</tr>
<tr>
<td>Presence of stroke</td>
<td>absence of CCA</td>
<td>aberrant ICA</td>
</tr>
</tbody>
</table>

However, stenotic lesions were present at the non-branching site in cases of significant carotid artery stenosis associated with non-bifurcating cervical carotid artery (Table 2). These findings suggest that the presence of a bifurcation or flow divider is not essential for the formation of atherosclerosis in the carotid artery. Table 2 shows that there is no difference in the pathological features of carotid plaque between normal vasculature and non-bifurcating cervical carotid artery. The carotid sinus is rich in baroreceptors. Baroreceptors are located in the anticipated site of the carotid bifurcation because this site of the carotid artery was demonstrated by vagal response to direct palpation during CEA. If agenesis of the proximal part of segment 1 occurs, the components of the carotid sinus including baroreceptors may remain in the anticipated site of the carotid bifurcation. We speculate that carotid plaques in patients with non-bifurcating cervical carotid artery would be formed as a result of the histological differences in this site and other local factors.

The present patient presented with amaurosis fugax. The ipsilateral ophthalmic artery was supplied by the middle meningeal artery due to carotid plaque because MR imaging showed asymptomatic ischemic lesions in the ipsilateral ICA territory. The findings of ultrasonography might support this inference because hypoechoic plaque indicating fragile components is associated with a high incidence of neurological events. Recent high resolution MR imaging could identify plaque constituents, such as the necrotic core and intraplaque hemorrhage.

Although high resolution MR imaging was not performed in the present case, the findings of other examinations could explain the mechanism of amaurosis.

CAS carries the risk of distal embolism during the perioperative period. Table 2 indicates that a stenotic lesion should be mostly located proximally to the ECA branches in non-bifurcating cervical carotid artery. Therefore, great care should be taken not to cause embolic complications in the territory of both the ICA and the ECA using CAS to treat patients with non-bifurcating cervical carotid artery. The collateral circulation must be investigated between the ECA and ophthalmic artery or ICA or vertebral artery. The present patient underwent CAS using two balloons for embolic protection of the ICA and middle meningeal artery because the ophthalmic artery was supplied by the middle meningeal artery, a branch of the internal maxillary artery. CAS can be safely employed for atherosclerotic stenosis associated with non-bifurcating cervical carotid artery provided preoperative assessment of collateral circulation is performed.

**Acknowledgments**

We would like to express our gratitude to Masaki Komiyama, MD of Osaka City General Hospital, Osaka, Japan for assisting with manuscript preparation.

**Conflicts of Interest Disclosure**

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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Address reprint requests to: Tetsuo Sasaki, MD. Interventional Neuroradiology Center, Aizawa Hospital, 2–5–1 Honjo, Matsumoto, Nagano 390–8510, Japan.

E-mail: sasakitetsu@gmail.com