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Carotid Endarterectomy for Vertebrobasilar Insufficiency Caused by Severe Stenosis of Primitive Hypoglossal Artery: A Technical Case Report and Literature Review

Masafumi SEGAWA,[1] Tomohiro INOUE,[1] Sho TSUNODA,[1] Ryuuichi NODA,[1] and Atsuya AKABANE[1]

[1]*Department of Neurosurgery, NTT Medical Center Tokyo, Shinagawa-ku, Tokyo, Japan*

Abstract

Persistent primitive hypoglossal artery (PHA) originating from the external carotid artery (ECA) is a rare anomaly. Reports of carotid endarterectomy (CEA) for ECA stenosis associated with this anomaly are even rarer. A 76-year-old woman presented to a medical clinic with a major complaint of refractory dizziness. Carotid ultrasound study suggested severe stenosis of the left cervical carotid bifurcation; therefore, she was referred to our department for a possible CEA. The imaging results indicated severe stenosis of the left carotid bifurcation and that the ECA was a PHA and the origin of the dominant vertebrobasilar artery (VBA). CEA was performed with the special caution of providing VBA collateral flow during clamping and preventing microembolisms during declamping of the ECA. Postoperative head magnetic resonance imaging revealed no new findings of cerebral infarction, and her dizziness disappeared. CEA associated with stenosis of the PHA as the origin of a dominant VBA was safely performed with an appropriate understanding of possible collateral pathways during cross-clamping.

Keywords: persistent primitive hypoglossal artery, arterial anomaly, carotid endarterectomy, occipital artery, vertebral artery

Introduction

Persistent primitive hypoglossal artery (PHA) is a carotid-basilar artery anastomosis that is a rare congenital arterial anomaly (estimated incidence of 0.02%-0.26%).[1)] PHA originating from the external carotid artery (ECA) is defined as "PHA type 2" and is even rarer.[2)] In these cases, intracranial aneurysms and stenosis of the carotid artery are known to be complicated, with some reports on their treatment.[3,4)] Carotid endarterectomy (CEA) associated with PHA type 2 has a higher risk of intraoperative cerebral and brainstem infarction compared with typical CEAs because prograde blood flow in the anterior and posterior circulations is simultaneously clamped. We describe a case of successful revascularization that was achieved through neuromonitoring, surgical technique, and edaravone in a patient with severe carotid bifurcation stenosis associated with PHA type 2.

Case Report

A 76-year-old woman with a major complaint of refractory dizziness was referred to our hospital by another hospital. Magnetic resonance (MR) imaging (MRI) revealed severe carotid bifurcation stenosis (Fig. 1A) and the branch of ECA, passing into the skull through the hypoglossal canal (Fig. 1B). Cerebral angiography revealed severe stenosis of the left carotid bifurcation and an arterial anomaly, the so-called PHA, originating from the ECA and anastomosing to the left vertebral artery (VA) through the left hypoglossal canal (Fig. 1C). Although the right VA was hypoplastic, inflow of the contrast medium was observed up to the basilar artery, and the left posterior communicating artery (PCOMA) was vaguely observed on angiography (Fig. 1D). In addition, collateral circulation from the left occipital artery (OA) to the left VA was noted (Fig. 2A). In our patient, the PHA was mainly responsible for the blood flow in the

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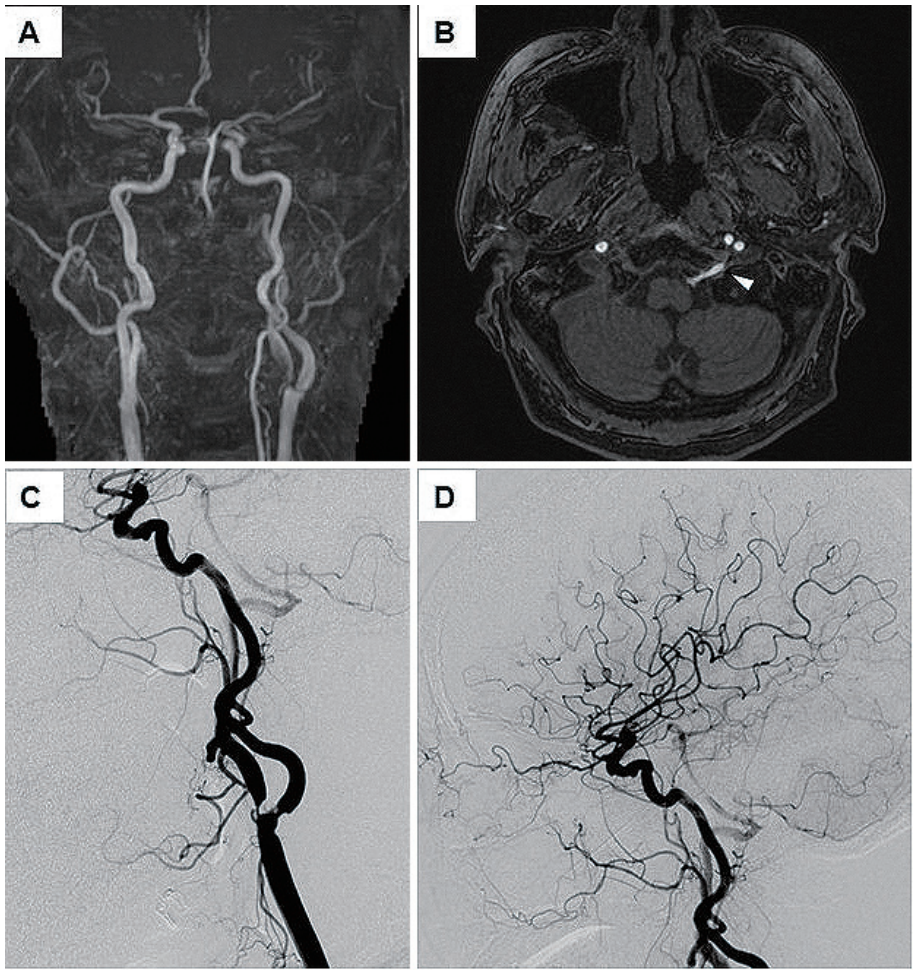
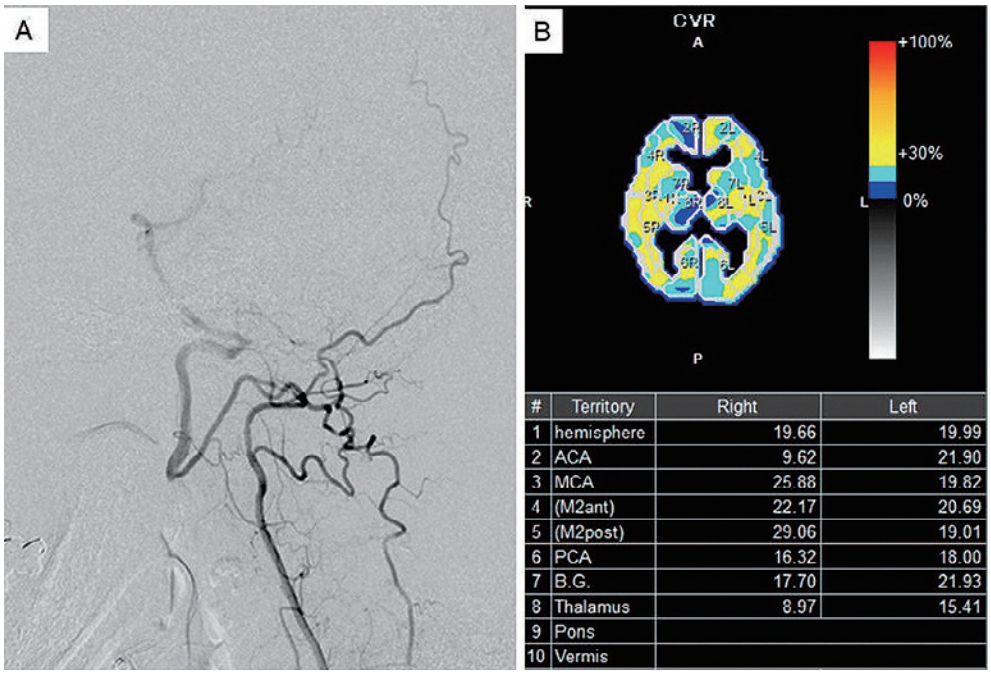


Fig. 1 (A) Initial magnetic resonance (MR) angiography on admission. (B) In this case, MR angiography revealed that the blood vessel is running inside the left hypoglossal canal, which was consistent with PHA (white arrow). (C) (D) Lateral view of the left common carotid artery (CCA) on a preoperative angiogram focused on the neck and head.

posterior circulation, but due to the presence of the right VA and left PCOMA, it was assumed that ischemic tolerance might be preserved by clamping the ECA branching from the PHA when an intraluminal shunt was inserted into the internal carotid artery (ICA) and common carotid artery (CCA). Preoperative N-isopropyl-p-(iodine-123)-iodoamphetamine (^{123}I-IMP)-single-photon emission computed tomography of acetazolamide loading revealed a decrease in cerebrovascular reactivity in the posterior cerebral artery (PCA) region compared with that in the bilateral middle cerebral artery (MCA) region (right MCA: 25.88%, left MCA: 19.82%; right PCA: 16.32%, left PCA: 18.00%) (Fig. 2B). Therefore, her dizziness might have been caused by vertebrobasilar insufficiency. As it was symptomatic, we decided to perform surgery. This patient required reliable patency of the ECA and ICA. Carotid stenting has a higher risk in postoperative ECA stenosis and occlusion; therefore, we decided to perform CEA in this patient.[5] Transcranial near-infrared spectroscopy (NIRS), both-upper-limb somatosensory evoked potential (SSEP), and auditory brain stem response (ABR) evaluations were performed with the patient under general anesthesia to monitor cerebral and brainstem ischemia. The SSEPs and ABR were measured every 5 min to monitor the latency prolongation and amplitude reduction. The back-plate was raised to 15° and fixed in the cervical extension position. An S-shaped skin incision was made along the anterior edge of the sternocleidomastoid muscle, the sternocleidomastoid muscle was expanded posterolaterally, and the ansa cervicalis, CCA, and internal jugular vein were proximally identified. To secure the CCA, the carotid sheath was opened. We expanded the internal jugular vein posterolaterally while chasing the ansa cervicalis to the distal point where it joins the hypoglossal nerve. We followed the CCA to secure the ECA and superior thyroid artery (STA) (Fig. 3A). Subsequently, the ICA was secured, and a bolus of heparin (5000 U) was administered before ICA clamping. In addition, 30 mg of edaravone (Radicut; Mitsubishi Pharma Corp., Tokyo, Japan) was administered *via* drip infusion for 30 min before ICA clamping. The STA, CCA,



#	Territory	Right	Left
1	hemisphere	19.66	19.99
2	ACA	9.62	21.90
3	MCA	25.88	19.82
4	(M2ant)	22.17	20.69
5	(M2post)	29.06	19.01
6	PCA	16.32	18.00
7	B.G.	17.70	21.93
8	Thalamus	8.97	15.41
9	Pons		
10	Vermis		

Fig. 2 (A) Lateral view of the left vertebral artery on a preoperative angiogram. (B) Cerebrovascular reactivity (CVR) in preoperative N-isopropyl-p-(iodine-123)-iodoamphetamine (^{123}I-IMP)-single photon emission computed tomography (SPECT) examination of acetazolamide loading.

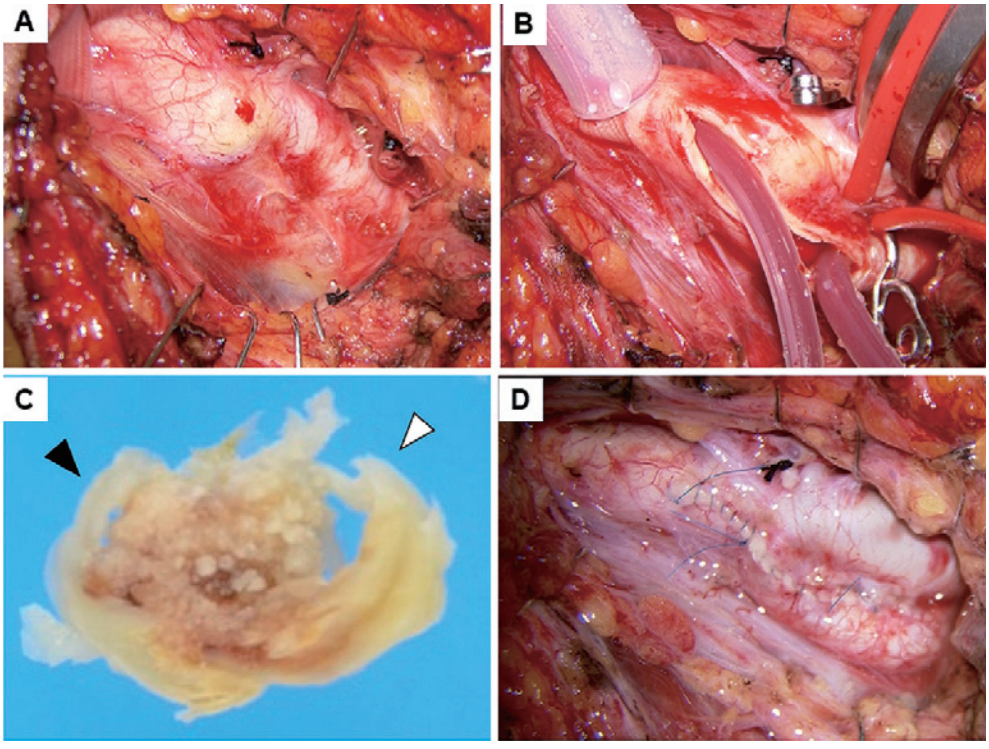


Fig. 3 Operative view. (A) The left common carotid artery (CCA), external carotid artery, and superior thyroid artery were secured after carotid sheath opening. (B) An intraluminal shunt was placed into the left CCA and internal carotid artery. (C) Cross section of the carotid bifurcation plaque (white arrow: internal carotid artery, black arrow: external carotid artery). (D) Final view of the carotid endarterectomy.

ECA, and ICA were temporarily clamped in order. An arterial incision was made, a T-shaped intraluminal shunt was inserted into the ICA and CCA (Fig. 3B), and the thrombus intima was removed.

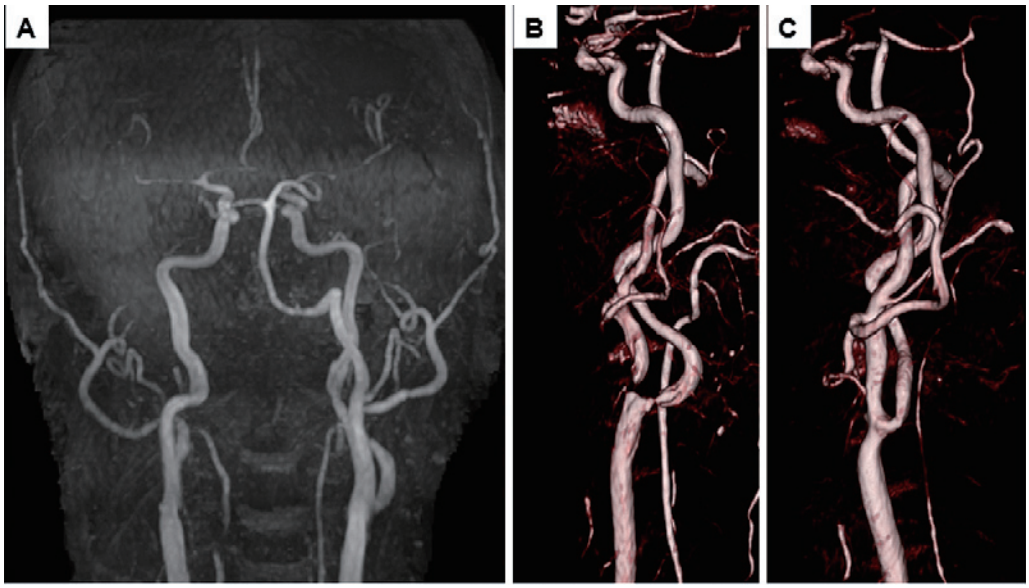


Fig. 4 (A) Postoperative magnetic resonance (MR) angiography. (B) Preoperative MR angiography of the neck. (C) Postoperative MR angiography of the neck.

Table 1 Reported cases of PHA from the ECA associated with CEA

Author	Age	Side	Symptom	OA–VA anastomosis	Contralateral VA	Treatment
Welten et al. 1988	68	Lt	Gait disturbance	None	Hypoplastic	Lt. CEA
Nanto et al. 2012	63	Lt	Asymptomatic	None	Hypoplastic	Lt. CEA
Present case. 2021	76	Lt	Dizziness	From OA toward VA	Hypoplastic	Lt. CEA

ECA: external carotid artery, CEA: carotid endarterectomy, OA: occipital artery, PHA: persistent hypoglossal artery, VA: vertebral artery.

Macroscopically, a high degree of calcification on the arterial wall was observed, which was protruding into the lumen at the bifurcation of the ICA and ECA (Fig. 3C). The temporary clamp was released by performing a procedure different from the usual one, and the artery was sutured with 6-0 Prolene (Fig. 3D). The actual surgical procedure is demonstrated below and can also be checked in the supplemental online video (Video 1). In SSEPs and ABR, no particular change was observed during the surgery because of the clamping arteries. Postoperative head MRI revealed no new findings of cerebral and brainstem infarction; an improvement in the depiction of the VA from the PHA was noted (Fig. 4A). Three-dimensional reconstruction of the cervical MR angiography before and after surgery revealed improvement in the stenosis of the carotid bifurcation and obscuration of the depiction of the left VA. This finding indicated that the improvement of anterograde blood flow from the OA reduced the blood flow from the VA to the ECA *via* the OA, which was observed before surgery (Fig. 4 B, C). Postoperative arterial spin-labeling MR perfusion showed increased blood flow qualitatively in the posterior circulation compared with the preoperative flow. Her dizziness disappeared after surgery. She was discharged with a modified Rankin scale score of 0. At the 7-month postoperative follow-up, the patient remained symptom-free.

Discussion

We performed CEA to treat carotid bifurcation stenosis associated with PHA type 2 and achieved a good clinical outcome for two reasons: preoperative understanding of 1) PHA embryology and anatomy and 2) surgical technique.

Embryology and anatomy

Padget et al. first reported primitive carotid-basilar anastomosis in 1948 and that early embryonic hindbrain receives blood flow from four blood vessels, namely, the trigeminal, otic, and hypoglossal nerves and the proatlantal artery.[6] These vessels usually involute rapidly with the development of the PCOMA and VA but sometimes remain in adults. PHA running from the ECA is an extremely rare variant of the PHA, and Uchino et al. named this variant "PHA type 2." The hypoglossal branch of the ascending pharyngeal artery from the ECA, passing into the skull through the hypoglossal canal, is considered a remnant of the PHA.[2,7] In the PHA, the contralateral VA and PCOMA

are only present in one-third of cases. In the setting of the PHA, at least one vertebral or PCOMA is hypoplastic or atretic in 86% and 94% of cases, respectively, and both are hypoplastic or atretic in 28% and 78%, respectively.[8] Most cases of carotid artery stenosis associated with PHA might have low ischemic tolerance when cervical blood vessels are clamped during CEA.

This case also involved OA-VA anastomosis. The frequency of observing this anastomosis on cerebral angiography is 8/1000 cases and is more often recognized in patients with vascular occlusive disease.[9] Severe stenosis of the ECA may generate a pressure gradient from the VA toward the ECA *via* the OA. To date, 11 cases of patients with PHA arising from the ECA have been reported,[2,3,10-16] and two of them underwent CEA (Table 1).[13,16] To the best of our knowledge, this case is the first to report a PHA running from the ECA and an OA-VA anastomosis associated with CEA.

Surgical technique

During CEA, SSEPs on the upper limb opposite the CEA and bilateral transcranial NIRS are typically used for neuromonitoring performed routinely at our institution; however, in this case, SSEPs on the left-upper limb and ABR were added to monitor brainstem ischemia.[17] Assuming that marked deterioration of the intraoperative ABR and left SSEPs might occur, we exposed the ECA enough distally so that we could replace the distal shunt tube from the ICA into ECA. Ischemia-reperfusion injury due to temporary clamping of the vertebrobasilar artery may be detected by ABR and SSEP on the ipsilateral side of the CEA, which is not commonly used. Edaravone, a novel free-radical scavenger, was used before the ICA and ECA clamping to prevent reperfusion injury.[18] When securing the cervical blood vessel, the collateral circulation was preserved by clamping the site just proximal to the OA. In almost all autopsy cases, OA-VA anastomosis is observed,[9] and blood flow to the posterior circulation may be expected by performing CEA without clamping the OA in PHA type 2.

We changed the order of declamping blood vessels from that followed in the standard procedure. After the removal of the intraluminal shunt, the debris is released to the STA to prevent intraoperative microembolism. In the standard procedure, the ICA is first declamped for a few seconds. Then, the STA, ECA, and CCA are declamped in order. After 30 s, the ICA is declamped. In the present case, the STA was first declamped. Second, the CCA was declamped, and a Doppler ultrasound flow meter was used to measure the STA prograde flow. Third, the ICA was declamped temporarily for 10 s, and the ECA was then declamped to allow debris to flow out into the STA. Finally, the CCA and ICA were declamped in order. In postoperative imaging, no new cerebral or brainstem infarction was observed.

Conclusion

PHA running from the ECA is rarely observed. When performing CEA, comprehensive understanding of the anatomy and hemodynamics by performing preoperative cerebral angiography is necessary. Intraoperative neuromonitoring, the order of clamping arteries, and the use of edaravone might have helped us safely perform CEA in this patient. Insufficient development of the contralateral VA and PCOMA might have maintained the ischemic tolerance development of the collateral circulation, such as *via* the OA-VA anastomosis.

Supplementary Material

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Acknowledgments

None.

Ethics Approval and Consent to Participate

Formal approval from a review board was not required for the systemic review part of the study. For the case report, patient consent was obtained.

Conflicts of Interest Disclosure

The authors have no potential conflicts of interest related to this report.

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Corresponding author: Masafumi Segawa, MD.
NTT Medical Center Tokyo, 5-9-22 Higashigotanda, Shinagawa-ku, Tokyo 141-0022, Japan.
e-mail: segawa.neuro.m@outlook.jp