Carotid Artery Stenting for Symptomatic Stenosis of the Cervical Carotid Artery Associated with Persistent Primitive Hypoglossal Artery: A Case Report

Shogo Kaku,1 Kengo Nishimura,1 Michiyasu Fuga,2 Masahide Watanabe,2 Tetsuaki Iwamoto,2 and Yuichi Murayama1

Objective: A case of symptomatic cervical carotid artery stenosis with persistent primitive hypoglossal artery (PPHA) treated by carotid artery stenting (CAS) with appropriate embolic protection is reported.

Case Presentation: The patient was a 65-year-old male presenting with left hemiplegia. MRI revealed infarction in the right cerebral hemisphere, and cerebral angiography demonstrated stenosis affecting the proximal segment of the internal carotid artery (ICA), proximal to the origin of the PPHA. Since blood flow was observed from the PPHA to the ICA during simultaneous obstruction of the common and external carotid arteries, a filter protection device was placed in the ICA along with proximal protection, and CAS could be performed without complications.

Conclusion: In performing CAS for symptomatic stenosis of the cervical carotid artery with PPHA, it is considered important to select appropriate embolic protection based on the evaluation of the direction of the blood flow of the ICA and PPHA under balloon occlusion conditions.

Keywords ▶ carotid artery stenting, persistent primitive hypoglossal artery, cervical internal carotid artery stenosis, endovascular surgery

Introduction

Persistent primitive hypoglossal artery (PPHA) is a very rare condition with an incidence of 0.02%–0.26%.1,2) We encountered a patient with symptomatic cervical carotid artery stenosis with coexisting PPHA and treated the condition by carotid artery stenting (CAS). How to achieve embolic protection in such patients with unusual and complex hemodynamics is a major problem. We could perform CAS safely without complications by carrying out angiography of the common carotid artery (CCA) during simultaneous occlusion of the CCA and external carotid artery (ECA), following the direction of washout of the contrast agent that reached the PPHA, and selecting an appropriate method for embolic protection. This case is reported with a review of the literature.

Case Presentation

A 65-year-old male with a history of hypertension awoke with left hemiplegia and he was urgently transported to our hospital.

On arrival, he was awake and alert but showed left hemiparesis (manual muscle test; MMT 4/V). Electrocardiography, echocardiography, and chest contrast-enhanced CT evaluation showed no clear indications for arrhythmia, persistent foramen ovale, or atherosclerotic or dissecting lesion of the aortic arch.

Radiographic findings

Head and neck MRI/MRA: Head MRI (diffusion-weighted imaging) on admission showed hyperintensities in the right precentral gyrus, and MRA showed right cervical internal carotid artery (ICA) stenosis associated with a persistent...
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revascularization. CAS was selected as the method of choice according to the following therapeutic strategy:

1) For the early clinical detection of intra-procedural complications, particularly embolic, the procedure will be performed under local anesthesia. If symptoms of ischemia appear, it can be proceeded with sedation of the patient and discontinuation of the procedure.

2) Proximal protection will be implemented since the possibility of vulnerable plaque accompanied with partial
ulceration could not be excluded despite the low degree of stenosis.

3) Distal protection should be attempted since there was concern that the superior thyroid artery might not be completely occluded if proximal protection is applied.

4) The direction of washout of the contrast agent that reached the ICA and PPHA has to be evaluated by performing CCA angiography with simultaneous obstruction of the CCA and ECA to determine the site of placement of the distal protection device.

**Endovascular treatment (CAS)**

According to the above-mentioned therapeutic strategy, a 9 Fr long sheath was inserted into the right femoral artery under local anesthesia, a 4 Fr short sheath was inserted into the left femoral vein, they were connected with an Optimo chamber (Tokai Medical Products, Aichi, Japan), and modified Parodi’s technique was carried out. After the activated clotting time was adjusted to ≥300 seconds by systemic heparinization, a 9 Fr OPTIMO 90 cm (Tokai Medical Products) was placed in the CCA, and a Carotid Guardwire PS (Medtronic, Minneapolis, MN, USA) was placed in the ECA. Then, both the CCA and ECA were obstructed by inflating the balloons, and angiography was performed. Since a reverse flow from the PPHA to the ICA was observed (Fig. 4A and 4B), a FilterWire EZ (Boston Scientific, Natick, MA, USA) was placed in the ICA (Fig. 4C). Next, pre-dilatation was made by inflating a Sterling MR (Boston Scientific) 3.5 mm × 20 mm at 6 atm for 30 seconds, debris was sufficiently aspirated, and the procedure was ended (devascularization time: 17 min) (Fig. 5). There were no postprocedural complications, and, after rehabilitation, the patient was discharged in a condition capable of unassisted ambulation.

**Discussion**

The primitive vascular anastomosis connecting the ICA with the vertebrobasilar artery is formed at the 4-mm embryonic stage, but regresses in the process of development of the vertebral and posterior communicating arteries, and usually disappears completely during the 14–18 mm embryonic stage, but if anastomotic vessels remain for some reason in this process, they result in persistent carotid-basilar anastomoses. The frequency of persistence is reported to be highest at 0.1%–0.2% for the trigeminal artery,3) followed by 0.02%–0.26% for the PPHA.1,2) Since the PPHA originates from the ICA, passes through the hypoglossal canal, and anastomoses with the vertebrobasilar system, 3D-CTA is considered useful for its diagnosis.4,5) In addition, as diagnostic criteria for PPHA, Lie et al.6) proposed 1) origin from the ICA at the C1–C3 level, 2) contrast-enhancement of the basilar artery only distally...
to the anastomosis of the abnormal vessel, 3) entry of the abnormal vessel into the cranium through the hypoglos-
sal canal, and 4) no contrast enhancement of the ipsilat-
eral posterior communicating artery. Since our patient
fulfilled all these diagnostic criteria on 3D-CTA and
angiography, we confirmed the condition as PPHA.
When treating cervical CAS associated with PPHA, the
choice between carotid endarterectomy (CEA) and CAS
remains a controversial issue. However, in the presence
of PPHA, the contralateral vertebral artery is often hypo-
plastic and the ipsilateral vertebral artery proximal to the
PPHA junction is often also hypoplastic or aplastic, so
it is necessary for CEA to place intra-operative shunts in
both the ICA and the PPHA. In addition, as the site of
stenosis is relatively high, that is, at the C1–C3 level,
CAS has been recently selected in some patients, as
we did in our patient too. We also selected local anesthe-
sia to allow early detection of symptoms due to intraop-
erative embolic complications or occlusion intolerance.
However, if symptoms of ischemia should occur, it is
considered desirable to sedate the patient and end the
procedure as promptly as possible. We planned the use of
distal protection through the procedure because of the
possibility of ulcerated and vulnerable plaque (although
stenosis was not advanced) and the possible inability to

Table 1: Summary of the previous reports of CAS for ICA stenosis associated with PPHA

<table>
<thead>
<tr>
<th>Case report</th>
<th>Age/Sex</th>
<th>Stenotic lesion</th>
<th>Distal flow control</th>
<th>Proximal flow control</th>
<th>Anesthesia</th>
<th>Occlusion time (min)</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kanazawa et al.9)</td>
<td>68/M</td>
<td>ICA stenosis (proximal to the origin of the PPHA)</td>
<td>Balloon</td>
<td>Reversal</td>
<td>General</td>
<td>8</td>
<td>None</td>
</tr>
<tr>
<td>Nii et al.10)</td>
<td>62/M</td>
<td>ICA stenosis (proximal to the origin of the PPHA)</td>
<td>None</td>
<td>None</td>
<td>General</td>
<td>11</td>
<td>None</td>
</tr>
<tr>
<td>Silva et al.11)</td>
<td>63/F</td>
<td>Extension of the ICA stenosis to the origin of the PPHA</td>
<td>Balloon</td>
<td>None</td>
<td>General</td>
<td>ND</td>
<td>None</td>
</tr>
<tr>
<td>Eller et al.12)</td>
<td>65/ND</td>
<td>ICA and PPHA stenosis</td>
<td>None</td>
<td>Reversal</td>
<td>Local</td>
<td>ND</td>
<td>None</td>
</tr>
<tr>
<td>Zhang et al.13)</td>
<td>47/M</td>
<td>Extension of the ICA stenosis to the origin of the PPHA</td>
<td>None</td>
<td>Blockade</td>
<td>General</td>
<td>13</td>
<td>None</td>
</tr>
<tr>
<td>Ryu et al.14)</td>
<td>60/F</td>
<td>ICA stenosis (proximal to the origin of the PPHA)</td>
<td>Balloon</td>
<td>Blockade</td>
<td>Local</td>
<td>18</td>
<td>None</td>
</tr>
<tr>
<td>Murai et al.18)</td>
<td>77/M</td>
<td>ICA stenosis (proximal to the origin of the PPHA)</td>
<td>Filter</td>
<td>Blockade</td>
<td>General</td>
<td>11</td>
<td>None</td>
</tr>
<tr>
<td>Current case</td>
<td>65/M</td>
<td>ICA stenosis (distal to the CCA)</td>
<td>Filter</td>
<td>Reversal</td>
<td>Local</td>
<td>17</td>
<td>None</td>
</tr>
</tbody>
</table>

CAS: carotid artery stenting; CCA: common carotid artery; F: female; ICA: internal carotid artery; M: male; ND: not described; PPHA: persistent primitive hypoglossal artery
exclude the superior thyroid artery by the proximal balloon protection. In addition, following the report by Murai et al., we performed CCA angiography by occluding both the CCA and ECA and confirmed the direction of washout of the contrast agent from PPHA (retrograde flow) to ICA (anterograde flow).

To our knowledge, CAS has been performed for symptomatic cervical carotid artery stenosis complicated by PPHA in only seven cases until now (Table 1). Common observations in these seven cases are as follows: 1) The site of placement of embolic protection and the device should be selected depending on the site of the lesion (stenosis), 2) a flow reversal system should be installed after controlling the proximal flow, and 3) while the procedure can be performed safely without body movements under general anesthesia, local anesthesia may also be an option as it is advantageous for the evaluation of embolic complications and occlusion intolerance.

In performing CAS for cervical carotid artery stenosis complicated by persistent primitive arteries such as PPHA, it is considered necessary to formulate a tailor-made therapeutic strategy for each patient unlike the standard procedure due to the specific individual anatomical characteristics and the existing flow pattern.

| Conclusion |

In performing CAS for symptomatic cervical carotid artery stenosis complicated by PPHA, it is considered important to select appropriate embolic protection after establishing the site of stenosis and the flow pattern of the ICA and PPHA by angiography with occlusion of the ICA and ECA.

| Disclosure Statement |

There are no conflicts of interest to disclose regarding this paper.

| References |