Carotid-Cavernous Fistula Caused by Rupture of Persistent Primitive Trigeminal Artery Trunk Aneurysm
—Case Report—

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Abstract
A 60-year-old female presented with a carotid-cavernous fistula (CCF) manifesting as left abducens nerve palsy. Left internal carotid digital subtraction angiography showed a persistent primitive trigeminal artery (PPTA) near the CCF. Super-selective angiography showed direct shunt flow between the PPTA trunk aneurysm and the left cavernous sinus. The aneurysm was successfully occluded with detachable coils. The CCF disappeared and the PPTA was preserved. The abducens nerve paralysis had disappeared 6 months later. CCF caused by a PPTA trunk aneurysm is extremely rare. We speculate that the PPTA trunk aneurysm formed and then ruptured due to hemodynamic stress caused by hypoplasia of the basilar artery.

Key words: carotid-cavernous fistula, persistent primitive trigeminal artery, coil embolization

Introduction
Persistent primitive trigeminal artery (PPTA) is an embryonic communication between the basilar and internal carotid arteries,26) which usually arises from the presellar internal carotid artery as it exits in the carotid canal and enters the cavernous sinus, then extends posteriorly to join the distal third of the basilar artery, usually between the origins of the superior and anterior inferior cerebellar arteries. The incidence of PPTA is reported to be between 0.1% and 0.6% based on conventional angiography or magnetic resonance (MR) imaging.25) The presence of this artery is considered to indicate a defect in cerebrovascular development and may be associated with other vascular anomalies such as arteriovenous malformations or aneurysms.11) However, an aneurysm of the PPTA is rare. Rupture of a PPTA aneurysm can result in subarachnoid hemorrhage or carotid-cavernous fistula (CCF).22) Such CCFs are extremely rare. We describe a case of CCF caused by a PPTA trunk aneurysm successfully treated by intra-aneurysmal coil embolization.

Case Report
A 60-year-old female presented with a 6-month history of diplopia caused by left abducens nerve palsy but no history of head trauma. MR imaging revealed dilated cavernous sinus and cortical veins (Fig. 1). Single photon emission computed tomography (SPECT) showed hypoperfusion of the left frontal lobe (Fig. 2A). Left carotid digital subtraction angiography showed direct flow through a CCF into the cortical veins, and a PPTA arising from the carotid artery running to the basilar artery (Fig. 3A). Vertebral angiography performed to assess left neck common carotid artery compression (Alcock test) showed retrograde filling of the PPTA and CCF (Fig. 3B). Three-dimensional digital subtraction angiography revealed an aneurysm at the curved PPTA trunk (Fig. 4).

A micro-catheter (Excelsior 1018; Boston Scientific, Natick, Massachusetts, USA) was guided into the origin of the PPTA from the left internal carotid artery, and selective injection of contrast medium demonstrated direct flow into the cavernous sinus (Fig. 3C, D). The catheter was advanced further along the PPTA and the contrast injection repeated, allowing identification of an aneurysm

Fig. 1 Three-dimensional time-of-flight magnetic resonance image showing the dilated left cavernous sinus and cortical vein.
Fig. 2 Technetium-99m-ethylcysteinate dimer single photon emission computed tomography scans, before treatment (A) showing diffuse hypoperfusion in the left frontal lobe, and after treatment (B) demonstrating improved cerebral blood flow in the left frontal lobe.

arising from the anterior wall of the curved PPTA trunk, which was drained into the cavernous sinus (Fig. 3E). The aneurysm had a regular spherical shape, was approximately 6 mm in diameter, and had a relatively narrow neck. An attempt to pass the catheter through the fistula failed because the fistula was apparently too small. We decided to perform intra-aneurysm embolization with electrical detachable coils. After insertion of 6 Guglielmi detachable coils (total 55 cm; Boston Scientific), the aneurysm was successfully occluded, and the CCF completely disappeared with preservation of the PPTA (Fig. 5).

The patient recovered well, and the abducens nerve paralysis showed gradual recovery, with complete resolution 6 months after the procedure. Postoperative SPECT revealed improvement of the hypoperfusion in the left frontal lobe (Fig. 2B). Postoperative MR angiography showed narrowing of the basilar artery and the posterior circulation mainly depended on the PPTA (Fig. 6).

Discussion

Only 19 cases of trigeminal carotid fistula have been reported in the English literature, and 13 of these cases were non-traumatic (spontaneous) CCFs (Table 1). The locations of the fistula were the internal carotid artery-PPTA junction in 13 cases, and the PPTA trunk in 6. One case presented with intracerebral hemorrhage, but the symptoms in others were not distinctive and included bruits, chemosis, proptosis, diplopia, retro-orbital pain, and so on. The treatment methods included ligation or direct surgery in 3 cases, detachable balloons in 8, and coil embolization in 7. This disorder has been treated mainly by endovascular approaches since the 1990s. The present case of CCF caused by a PPTA trunk aneurysm was treated with intra-aneurysmal coiling. The 14 cases described as spontaneous CCFs, including our present...
Fig. 5 Digital subtraction angiograms performed during intra-aneurysmal embolization just after insertions of the first coil (A), the third coil (B), and the sixth (last) coil (C). The aneurysm was successfully occluded, and the carotid-cavernous fistula completely disappeared with preservation of the persistent primitive trigeminal artery (D).

Fig. 6 Pre- (A) and postoperative (B) magnetic resonance angiograms showing disappearance of the carotid-cavernous fistula and preservation of the persistent primitive trigeminal artery. The basilar artery seems to be hypoplastic.

case, may have been caused by rupture of a PPTA aneurysm, although aneurysms were detected on angiography in only 4 cases. The etiologies may be similar to those of CCFs caused by rupture of a cavernous aneurysm (Barrow type AII).11

Generally, the first choice of treatment for direct CCF is considered to be detachable balloon embolization.19 The transarterial approach is recommended for balloon embolization, and also usually recommended for coil embolization except in cases of Ehlers-Danlos syndrome with fragile arterial walls.13,18 Transarterial intra-aneurysmal embolization often effectively occludes the fistula.23 Coiling of the aneurysm with balloon protection was reported recently, and apparently provided favorable results in terms of fistula closure and precise preservation of the patency of the parent artery.15 Transvenous cavernous sinus packing with detachable coils may require a large volume of coils and carries the risk of ocular symptom exacerbation,15,24 but is considered a good choice if a transarterial approach is difficult.18 A major disadvantage of coiling is the high cost. Thus, from the standpoint of cost-effectiveness, aneurysm coiling might be preferable to sinus packing.23

The strategies we considered before treating this patient were embolization with a detachable balloon, trapping of the PPTA with coils, trans-arterial trans-fistula sinus packing with coils, trans-venous sinus packing with coils, and intra-aneurysmal coil embolization. Balloon embolization would have been too difficult because the parent artery was extremely tortuous and the fistula was quite small. Furthermore, detachable balloons are difficult to obtain commercially in Japan.18 PPTA is often associated with hypoplasia of the basilar artery, so trapping of the PPTA would have been technically feasible but not desirable in this case. The trans-arterial trans-fistula approach was also difficult due to the small fistula. Since transvenous sinus packing would require a large volume of coils, intra-aneurysmal embolization was the simplest treatment strategy available.

Super-selective angiography and aneurysmography facilitated our understanding of the morphology of the aneurysm. Vertebral angiography with ipsilateral carotid compression (Alcock test) was also useful for ascertaining the relationship between the PPTA and the fistula. Fortunately, the aneurysm was easily treated. The aneurysm was spherical and had a narrow neck, so we were able to occlude the fistula without difficulty. The internal carotid artery and the PPTA were preserved. If the neck had been wide, we would have adopted a balloon assist technique.29 We maintained the posterior circulatory blood flow, and SPECT revealed improved cerebral blood flow at the area of venous reflux. The findings of three-dimensional digital subtraction angiography were complicated, but this diagnostic procedure was very useful for measuring the diameter of the aneurysm at the time of selecting the first coil.

Various etiologies for aneurysms arising from the PPTA have been suggested, including dysplasia of the PPTA wall and hemodynamic stress on the PPTA.27 There is no histological evidence supporting the hypothesis that pathological anomalies develop in the PPTA wall. Based on a review of 261 cases with PPTA reported between 1950 and 2003, 39 cases were identified with PPTA aneurysms.20 These aneurysms were located not only at the arterial junction but also on the trunk of the PPTA, and were not always associated with connective tissue disorders such as
Table 1  Summary of the 20 reported cases of trigeminal carotid fistula

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age (yrs), Sex</th>
<th>Symptoms</th>
<th>Course</th>
<th>Location</th>
<th>Treatment</th>
<th>Sacrifice of ICA</th>
<th>Sacrifice of PTA</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enomoto et al. (1977)</td>
<td>42, F</td>
<td>ocular disorder</td>
<td>spontaneous (An)</td>
<td>ICA-PPTA (rt)</td>
<td>ICA ligation</td>
<td>yes</td>
<td>no</td>
<td>cure, mild ophthalmoplegia</td>
</tr>
<tr>
<td>Charlin et al. (1982)</td>
<td>53, F</td>
<td>ocular disorder</td>
<td>spontaneous (An)</td>
<td>PPTA trunk (lt)</td>
<td>balloon</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Kerber and Manke (1983)</td>
<td>26, M</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>PPTA trunk (lt)</td>
<td>balloon</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Berger and Hosobuchi (1984)</td>
<td>51, F</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>ICA-PPTA (lt)</td>
<td>balloon and surgery</td>
<td>yes</td>
<td>yes</td>
<td>cure, transient abducens palsy</td>
</tr>
<tr>
<td>Flandroy et al. (1987)</td>
<td>35, M</td>
<td>ocular disorder</td>
<td>traumatic</td>
<td>ICA-PPTA (rt)</td>
<td>balloon</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Debrun et al. (1988)</td>
<td>30, M</td>
<td>ocular disorder</td>
<td>traumatic</td>
<td>ICA-PPTA (rt)</td>
<td>balloon</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Cheng and Wang (1990)</td>
<td>42, M</td>
<td>ocular disorder</td>
<td>traumatic</td>
<td>ICA-PPTA (rt)</td>
<td>ICA ligation and surgery</td>
<td>yes</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Guglielmi et al. (1990)</td>
<td>20, M</td>
<td>ocular disorder</td>
<td>traumatic</td>
<td>PPTA trunk (rt)</td>
<td>refused</td>
<td>—</td>
<td>—</td>
<td>cure</td>
</tr>
<tr>
<td>McKenzie et al. (1990)</td>
<td>36, M</td>
<td>ocular disorder</td>
<td>traumatic (An)</td>
<td>PPTA trunk (lt)</td>
<td>balloon</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Bernstein et al. (1998)</td>
<td>53, F</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>ICA-PPTA (rt)</td>
<td>coil (TV)</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Hurst et al. (1999)</td>
<td>62, F</td>
<td>bruit, proptosis, diplopia</td>
<td>spontaneous</td>
<td>ICA-PPTA (lt)</td>
<td>coil (TA sinus packing)</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Masaryk et al. (1999)</td>
<td>43, F</td>
<td>bruit, eye symptom</td>
<td>spontaneous</td>
<td>ICA-PPTA (rt)</td>
<td>balloon</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Oka et al. (2000)</td>
<td>58, F</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>ICA-PPTA (rt)</td>
<td>coil (TV)</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Cook et al. (2000)</td>
<td>83, F</td>
<td>diplopia</td>
<td>traumatic</td>
<td>PPTA trunk (rt)</td>
<td>balloon</td>
<td>no</td>
<td>yes</td>
<td>cure</td>
</tr>
<tr>
<td>Tokunaga et al. (2004)</td>
<td>61, F</td>
<td>intracerebral hemorrhage</td>
<td>spontaneous</td>
<td>PPTA trunk (lt)</td>
<td>coil (TV)</td>
<td>no</td>
<td>no</td>
<td>cure, hemiparesis, dysphasia</td>
</tr>
<tr>
<td>Chan et al. (2006)</td>
<td>50, F</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>ICA-PPTA (rt)</td>
<td>coil (TV)</td>
<td>no</td>
<td>no</td>
<td>cure, mild ophthalmoplegia</td>
</tr>
<tr>
<td>Geibprasert et al. (2009)</td>
<td>35, F</td>
<td>ocular disorder</td>
<td>spontaneous</td>
<td>ICA-PPTA (rt)</td>
<td>coil (TV) and glue (TA)</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Xin-Ya Qian et al. (2009)</td>
<td>62, F</td>
<td>eye pain, chemosis, ophthalmoplegia</td>
<td>spontaneous (An)</td>
<td>ICA-PPTA (rt)</td>
<td>coil (TV)</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
<tr>
<td>Present case</td>
<td>60, F</td>
<td>bruit, abducens palsy</td>
<td>spontaneous (An)</td>
<td>PPTA trunk (lt)</td>
<td>coil (intra-An)</td>
<td>no</td>
<td>no</td>
<td>cure</td>
</tr>
</tbody>
</table>


Ehlers-Danlos syndrome. The site of most reported PPTA aneurysms has been the curved trunk of the PPTA, suggesting that high hemodynamic stress initiates aneurysm formation. In our case, the aneurysm was also situated on the curved trunk of the PPTA. Postoperative MR angiography indicated hypoplasia of basilar artery. Therefore, the findings in this case supported hemodynamic stress as the cause of the PPTA.

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