Carotid-cavernous Fistula Associated with a Ruptured Persistent Primitive Trigeminal Artery Aneurysm: A Case Report and Review of Literature

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Abstract

Carotid-cavernous sinus fistula (CCF) caused by a ruptured aneurysm of the persistent primitive trigeminal artery (PPTA) is rarely reported. A 69-year-old woman presented with progressive ptosis and pulsating tinnitus. Vertebral angiography under flow control of the internal carotid artery revealed CCF associated with a ruptured PPTA-trunk aneurysm, and PPTA was divided into Saltzman type 2. Endovascular treatment was performed by coil embolization of the aneurysm and parent artery occlusion of the PPTA, preserving the basilar artery (BA) side of PPTA, without complications. In the case of ruptured aneurysms originating from the Saltzman type 2 PPTA trunk, parent artery occlusion of the PPTA might be a treatment option and preservation of the BA side of PPTA is necessary to avoid ischemic complication of pons.

Keywords: carotid-cavernous sinus fistula, persistent primitive trigeminal artery, parent artery occlusion, coil embolization

Introduction

Persistent primitive trigeminal artery (PPTA) is the most common persistent primitive carotid-basilar anastomosis, with an incidence of 0.1–0.6%.[1] Carotid-cavernous sinus fistula (CCF) associated with a ruptured aneurysm of the PPTA is rare.[1,2] We present a case of CCF caused by a ruptured PPTA aneurysm that was successfully treated by coil embolization of the aneurysm and PPTA itself. We also reviewed the literature and considered the optimal treatment.

Case Report

A 69-year-old woman suddenly presented with pulsatile tinnitus and ptosis three months previously. Her symptoms worsened during the last two weeks, and she was referred to our institute. She had neither a history of head trauma nor intracranial surgery. Physical examination revealed left oculomotor nerve palsy, conjunctival hyperemia, and high-pitched bruit over her left eye and mastoid process. Her left intraocular pressure was slightly elevated. Magnetic resonance angiography (MRA) showed a vascular abnormality in the bilateral cavernous sinus (CS) and enlarged superior ophthalmic vein. Left internal carotid angiography and vertebral angiography revealed a high-flow shunt directly from the left PPTA to the left CS (Fig. 1A–1D).

The flow drained into the left superior ophthalmic vein, inferior petrosal sinus, contralateral CS, and bilateral superficial middle cerebral veins. Because of the high flow at that time, the aneurysm could not be detected, but the fistula point was suspected to be located at the PPTA trunk. Because of the progressive symptoms, urgent endovascular treatment was planned.

Dual antiplatelet therapy (75 mg/day of clopidogrel and 100 mg/day of aspirin) was administered 14 days before the surgery to avoid ischemic complications. Under general anesthesia, a 7F guiding
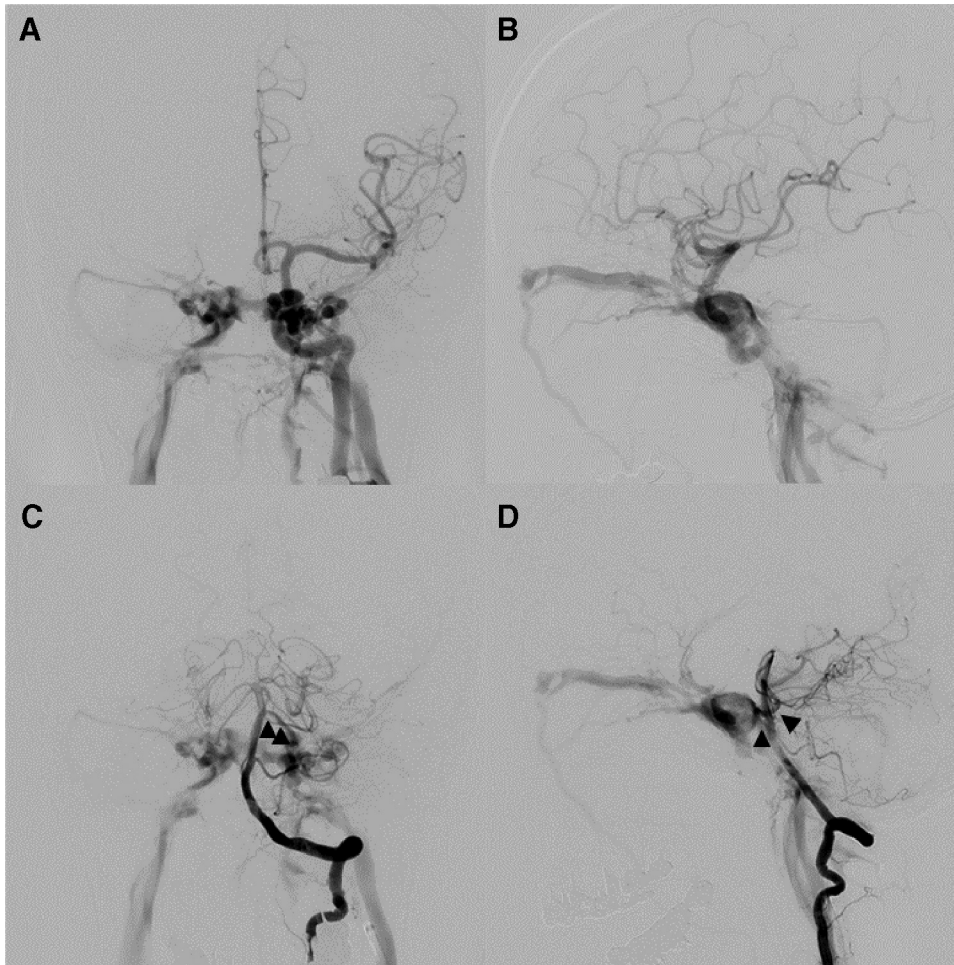
Fig. 1 (A and B) Left internal carotid angiography reveals high flow shunt from the left ICA and draining into the superior ophthalmic and facial vein, inferior petrosal sinus, contralateral CS, and bilateral superficial middle cerebral veins (A: anterior–posterior projection, B: lateral projection). **(C and D)** Left vertebral angiography reveals PPTA (black arrowheads) and high-flow shunt (C: anterior–posterior projection, D: lateral projection). CS: cavernous sinus, ICA: internal carotid artery, PPTA: persistent primitive trigeminal artery.

catheter (Roadmaster; Goodman CO., LTD., Aichi, Japan) was advanced into the left internal carotid artery (ICA) and a 6F guiding catheter (Envoy; Codman Neurovascular, Raynham, MA, USA) into the left vertebral artery via a transfemoral approach. While the ICA including the PPTA orifice was occluded by a balloon (Shouryu; Kaneka Medix Coop., Osaka, Japan), vertebral angiography revealed an aneurysm of the PPTA trunk (Fig. 2).

Thus, CCF caused by a ruptured PPTA aneurysm was diagnosed. A microcatheter (Excelsior SL-10; Stryker Neurovascular, Fremont, CA, USA) was advanced to the PPTA via the ICA. In addition, using a coaxial system consisting of a 4F intermediate catheter (Tactics; Technocrat, Aichi, Japan) and a microcatheter (SL-10), a microcatheter was advanced to the CS through the aneurysm via the

basilar artery (BA; Fig. 3A and 3B). First, the small component of CS and aneurysm, including fistulas, was occluded by seven detachable coils (Fig. 3C). Then, both BA and ICA sides of the PPTA trunk were occluded by 26 detachable coils, under temporary occlusion of ICA including the PPTA orifice by Shouryu for flow reduction and for protection of the coil protrusion to the ICA (Fig. 3D). PPTA of the BA side was preserved to avoid ischemic events due to the obliteration of perforators to the pons. These procedures resulted in complete obliteration of the shunt flow (Fig. 3E and 3F).

Her symptoms disappeared postoperatively without complications, and she was discharged 10 days after treatment. MRA examination showed no recurrence over two years after the treatment.
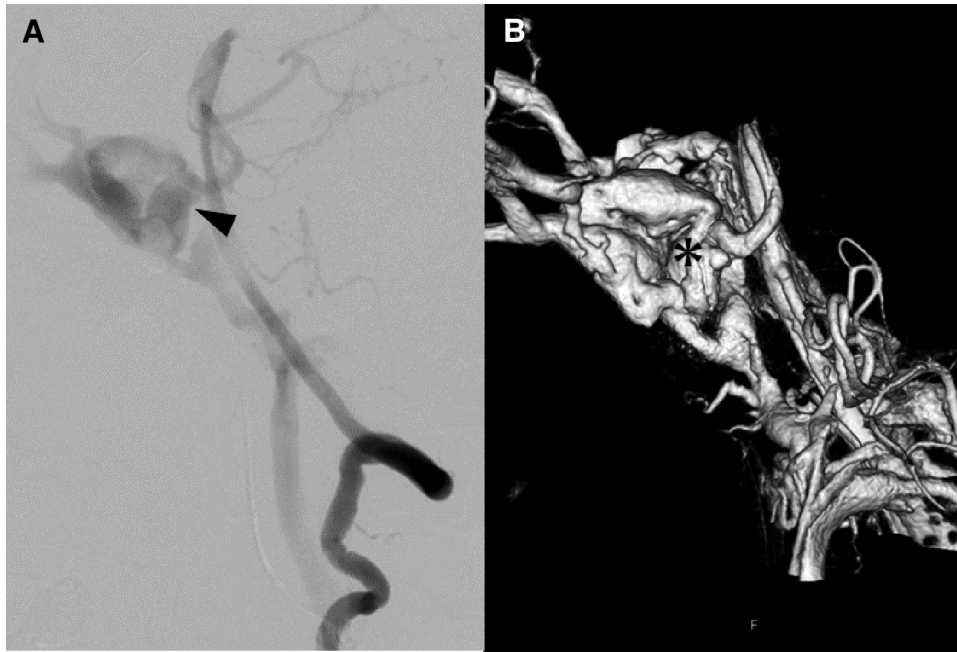
Fig. 2 (A and B) After ICA including the PPTA orifice is temporally occluded by the balloon, the aneurysm of the PPTA trunk is detected (A: arrowhead, B: asterisk in 3-dimensional digital subtraction angiography). ICA: internal carotid artery, PPTA: persistent primitive trigeminal artery.

Discussion

PPTA is the most common primitive artery, connecting the cavernous portion of the ICA and BA. CCF is a vascular shunt going directly from the carotid artery into the CS, and is caused by trauma (75%) and subsequently by spontaneous CCF (30%), including ruptured cavernous ICA aneurysms.[3] PPTA itself is rare, and the frequency of intracranial PPTA aneurysms is controversial (3.9–15.3%).[1,4] Limited data about ruptured PPTA aneurysms are available. Including our case, only 10 cases of spontaneous CCF associated with ruptured PPTA aneurysms were reported in the English literature (Table 1).[5–12]

In seven of the 10 cases, aneurysms originated from the PPTA trunk. In addition, most of the PPTA type was classified as Saltzman type 2, in which the BA proximal to the joint of the PPTA is well developed and there are posterior communicating arteries.[7,13] On the contrary, PPTA aneurysms associated with Saltzman types 1 and 3 were reported in 25% and 18%, respectively.[7] This was probably because the pathogenesis of PPTA aneurysms is possibly due to a combination of inherent congenital weakness of the middle layer of the PPTA artery and the hemodynamic stress caused by its anatomic location between the ICA and BA.[10,14] Ruptured PPTA aneurysms cannot be easily detected by routine

cerebral angiography because of the high-flow fistula from both the ICA and the BA to the CS. In our case, an aneurysm could not be detected by conventional cerebral angiography and could be detected after flow reduction of the ICA. Similarly, in the four reported cases, three-dimensional digital subtraction angiogram or flow reduction was needed to detect aneurysms (Table 1).

Endovascular treatment has recently become the first-line treatment. The prevalent treatment for CCF was detachable balloon treatment, but detachable balloons are not available in Japan. Transarterial embolization using detachable coils or liquid embolic agents is more recommended than transvenous embolization as embolization of the aneurysm sac by a transvenous approach is difficult, and transvenous CS packing with detachable coils may require a large number of coils and worse ocular symptoms.[15] Target transvenous and/or transarterial embolization is useful as a treatment option even in the CCF,[16,17] which prevents the complications of neurological symptoms caused by the CS packing and sacrifice of PPTA. However, this strategy is available in the case with small fistulas.[16] In our case, the precise dome and neck of the aneurysm were not identified, and the size of fistulas was not identified either. Thus, after the aneurysm including the fistulas was partially occluded, parent artery occlusion of the PPTA was performed.
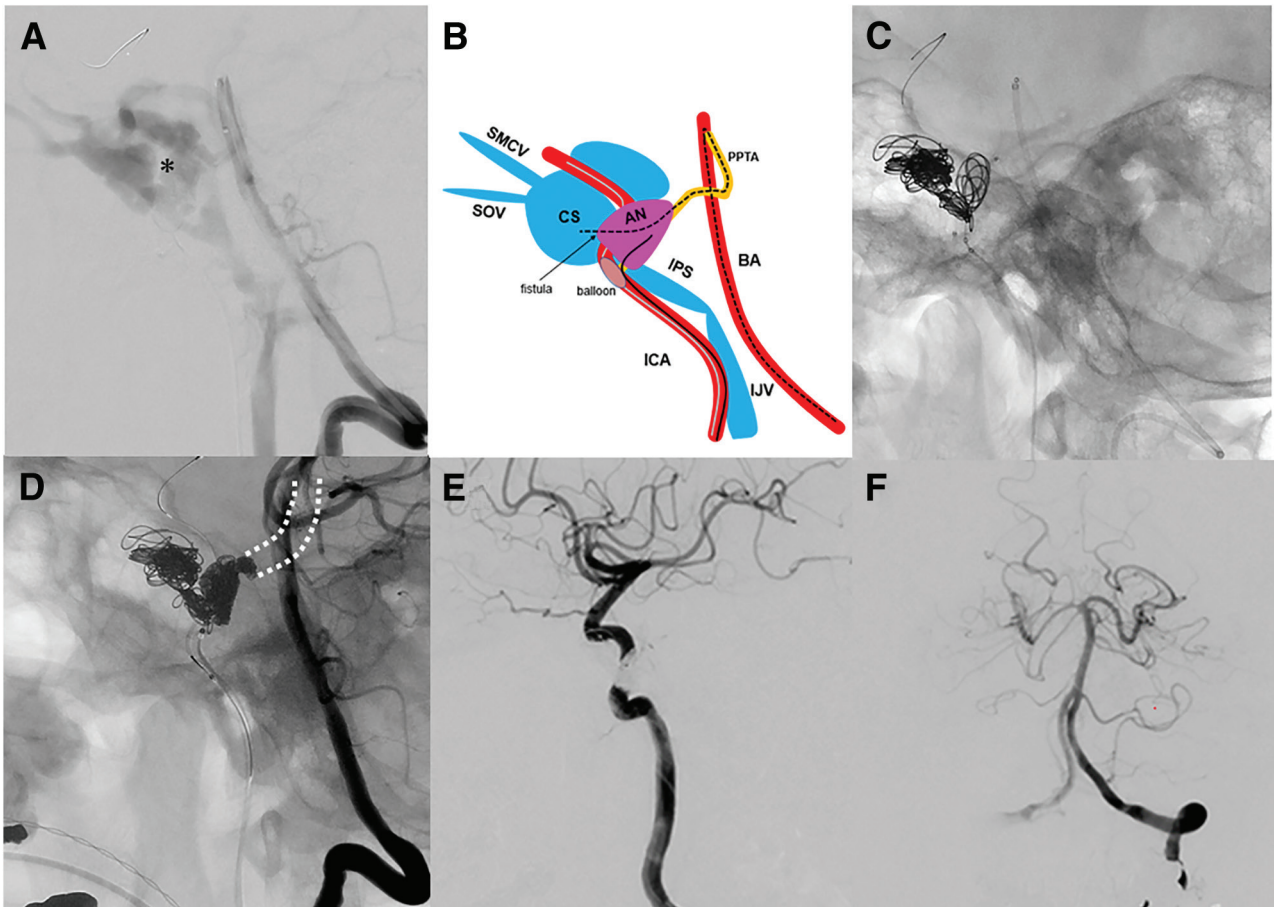
Fig. 3 (A) Microcatheters are advanced to the PPTA via the ICA and BA. Asterisk shows an AN of the PPTA trunk. (B) Schema of coil embolization using the double catheters. Black and black dot lines show microcatheters via the ICA and BA, respectively. (C) The small component of CS and aneurysm including fistulas is occluded by seven coils. (D) Parent artery occlusion of PPTA is performed. Of note, the PPTA of the BA side is preserved (white dot lines). (E and F) After parent artery occlusion of PPTA, shunt flow completely disappears (E: internal carotid angiography, F: vertebral angiography). AN: aneurysm, BA: basilar artery, CS: cavernous sinus, ICA: internal carotid artery, IJV: internal jugular vein, IPS: inferior petrosal sinus, PPTA: persistent primitive trigeminal artery, SMCV: superficial middle cerebral vein, SOV: superior ophthalmic vein.

In previous reports, including our case, coil embolization of the aneurysm was performed in six cases, and in four of those six cases, parent artery occlusion of the PPTA was performed. Of note, all those four cases, including our case, with parent artery occlusion of PPTA, classified as Saltzman type 2, had no ischemic complication. Therefore, in the cases of Saltzman type 2, parent artery occlusion of the PPTA including the aneurysms is one of the treatment options. In contrast, in the cases of Saltzman type 1, in which the BA proximal is hypoplastic and the posterior communicating arteries may be absent,[7] posterior circulation depends on the PPTA, and parent artery occlusion of the PPTA will result in a cerebral infarction in the posterior circulation. O'uchi and O'uchi[1] reported that PPTA

was divided into the medial and the lateral type; the medial type running superior to the abducens nerve had branches to the meningohypophyseal trunk, and the lateral type running inferolateral to the abducens nerve had perforators for the pons. In addition, the lateral type was 11 times as frequent as the medial type. Thus, even in the cases of parent artery occlusion of PPTA classified as Saltzman type 2, the occlusion of the BA side of the PPTA has the risk to cause ischemic complication of pons.

Conclusion

CCF associated with ruptured PPTA aneurysms is extremely rare. Although parent artery occlusion of the PPTA with the preservation of the BA side of

Table 1   Summary of the reported cases of carotid cavernous fistula from ruptured persistent primitive trigeminal artery aneurysms

Author (year)	Age/ sex	Symptoms	Location of aneurysm	Aneurysm detection	Saltzman classification	Treatment	ICA sacrifice	PPTA sacrifice	Outcome	Complications
Enomoto et al. (1977)[5]	42/F	Exophthalmos, diplopia	ICA–PPTA	Angiography	2	ICA ligation	Yes	No	Cure	Mild ophthalmoplegia
Charlin et al. (1982)*	53/F	Ocular disorder	PPTA trunk	NA	3 variant	Balloon	No	No	Cure	NA
Guglielmi et al. (1990)[6]	57/M	Diplopia, ophthalmoplegia	ICA–PPTA	No	2	Balloon	No	Yes	Cure	None
Qian et al. (2009)[12]	62/F	Chemosis, ophthalmoplegia	ICA–PPTA	No	2	Balloon	No	No	Cure	None
Liu et al. (2009)[11]	55/F	Diplopia, VI nerve palsy	PPTA trunk	Angiography	2	Coil	No	Yes	Cure	None
Kim et al. (2010)[10]	42/F	Diplopia, exophthalmos	PPTA trunk	Angiography (under flow reduction)	3 variant	Coil	No	No	Cure	None
Yoshida et al. (2011)[9]	60/F	Diplopia, VI nerve palsy	PPTA trunk	Angiography (3D)	1	Coil	No	No	Cure	NA
Fan et al. (2019)[8]	64/F	Diplopia, exophthalmos	PPTA trunk	Angiography (3D)	2	Coil and glue	No	No	Cure	None
Diana et al. (2019)[7]	61/F	Diplopia, exophthalmos	PPTA trunk	Angiography (3D)	2	Coil	No	Yes	Cure	None
Our case	69/F	Chemosis, III nerve palsy	PPTA trunk	Angiography (under flow reduction)	2	Coil	No	Yes	Cure	None

*We can only find the abstract. F: female, ICA: internal carotid artery, M: male, NA: not available, PPTA: persistent primitive trigeminal artery.

the PPTA might be a treatment option in the cases of Saltzman type 2, further case accumulation is warranted to determine the optimal therapy.

Conflicts of Interest Disclosure

All authors have no conflict of interest.

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