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Selective Transarterial Embolization for a Ruptured Persistent Trigeminal Artery Variant Aneurysm

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Abstract

We report a male patient with a ruptured persistent primitive trigeminal artery variant aneurysm that resulted in a fistula with the cavernous sinus. He presented with left conjunctival hyperemia and exophthalmos. Cerebral angiography revealed a left direct carotid-cavernous fistula; however, a balloon occlusion test determined that the source was actually a ruptured aneurysm located on the trunk of a persistent primitive trigeminal artery. Endovascular trapping of the persistent primitive trigeminal artery was performed, which resulted in fistula occlusion and symptom resolution.

Keywords: persistent primitive trigeminal artery, carotid-cavernous fistula, transarterial embolization, balloon occlusion test

Introduction

A persistent primitive trigeminal artery (PPTA) represents the remnant of a primitive anastomosis between the internal carotid artery (ICA) and vertebrobasilar system. Among the possible persistent primitive carotid-vertebrobasilar anastomoses, PPTA is the most cephalad and the most prevalent. Nonetheless, PPTA is rare with reported incidence rates ranging from 0.18% to 0.76%.[1,2)]

Persistent embryonic vessels may have structural wall defects.[3)] Reported prevalence rates of cerebral aneurysm in patients with PPTA range from 3% to 32%, which is quite wide owing to selection bias.[1,4)] Such aneurysms are typically located on the circle of Willis and at the ICA-PPTA bifurcation.[5)] Rupture of a PPTA aneurysm may cause subarachnoid hemorrhage or carotid-cavernous fistula (CCF). Cranial trauma is one cause of PPTA rupture.[6)] Today, endovascular treatment is preferred for PPTA aneurysms, although surgical clipping is also an option.

The Saltzman system classifies PPTAs into three types: type 1, 2, and 3 PPTAs. The type 3 PPTA, in which the PPTA terminates as cerebellar arteries instead of joining the BA,[7)] is frequently called a persistent trigeminal artery variant (PTAv). There are few reports of nontraumatic PTAv aneurysm rupture. We report a patient who developed a CCF after nontraumatic rupture of a PTAv aneurysm and underwent successful endovascular treatment.

Case Report

A 49-year-old man with left-sided conjunctival hyperemia and exophthalmos was referred to our institution for evaluation. Five months previously, he reported an episode of sudden severe headache, nausea, and pulsatile tinnitus. Physical examination revealed left exophthalmos, conjunctival hyperemia, eyelid swelling, ocular bruit, and oculomotor nerve palsy. Time-of-flight magnetic resonance angiography showed hyperintense signal in the left cavernous sinus (CS) and a contiguous aneurysmal dilation (Fig. 1A), left superior ophthalmic vein (SOV), and left inferior petro-

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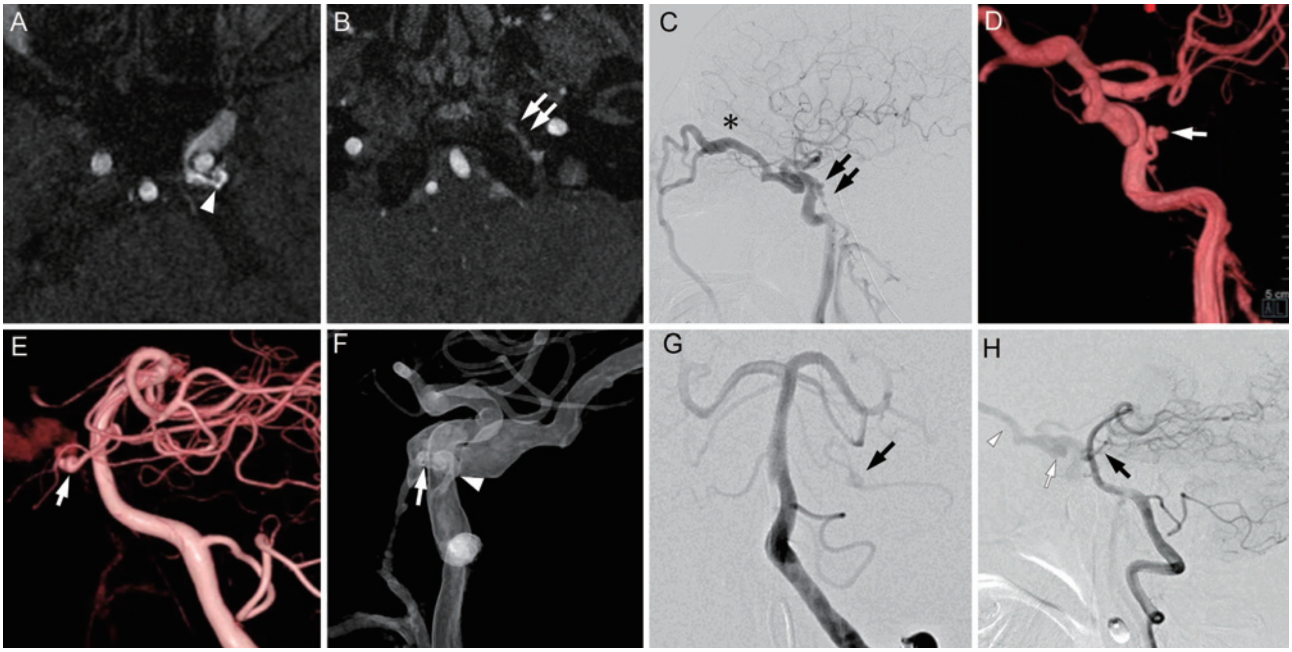

Fig. 1
A: Time-of-flight magnetic resonance angiography (axial cavernous sinus level) reveals flow signal in the left cavernous sinus connected from the aneurysmal dilation (white arrowhead).
B: Time-of-flight magnetic resonance angiography (axial medulla oblongata level) reveals flow signal in the left inferior petrosal sinus (white arrows).
C: Left internal carotid angiography (lateral view) shows shunted flow from the cavernous sinus into the left superior ophthalmic vein (asterisk) and left inferior petrosal sinus (black arrows).
D: Three-dimensional rotational left internal carotid angiography (lateral view) reveals an anomalous persistent primitive trigeminal artery (PPTA) (white arrow) arising from the C5 segment.
E: An aneurysm (white arrow) of the PPTA is also shown on three-dimensional rotational right vertebral artery angiography (lateral view).
F: The PPTA and (white arrowhead) its aneurysm are shown to be directly connected to the cavernous sinus (white arrow) on three-dimensional rotational left internal carotid angiography (right anterior oblique view).
G: Right vertebral angiography (anteroposterior view) during a balloon occlusion test of the left internal carotid artery emphasizes the PPTA with aneurysm connecting to the left anterior inferior cerebellar artery.
H: Right vertebral angiography (lateral view) during the balloon occlusion test shows the PPTA (black arrow) and its aneurysm (white arrow). Blood flows into the right superior ophthalmic vein (white arrowhead). The PPTA communicates with the distal region of the left anterior inferior cerebellar artery (black arrow).

sal sinus (IPS) (Fig. 1B). Left internal carotid angiography revealed shunted flow to the ipsilateral CS, which drained into the left SOV and left IPS (Fig. 1C) without cortical venous reflux. Three-dimensional rotational left ICA angiography revealed an anomalous artery arising from the C5 segment that had an aneurysmal dilation in its distal portion and connected to the left CS shunt (Fig. 1D). The same aneurysm and shunt were also demonstrated on right VA angiography (Fig. 1E). It was clearly shown that the aneurysm was directly connected to the CS by the right anterior oblique view of left ICA (Fig. 1F). The anomalous artery supplied the territory of the left anterior inferior cerebellar artery (AICA) (Fig. 1G).

Balloon occlusion of the proximal left ICA (Matas/Allcock maneuver) revealed good crossflow from the contralateral ICA via the anterior communicating artery and from the right VA via the PPTA and posterior communicating (Pcom) artery. Thirty minutes of occlusion resulted in no neurological symptoms or adverse event. Right VA angiography during the occlusion test emphasized the PPTA itself, its anastomosis with the BA, and the aneurysm of its trunk draining into the SOV (Fig. 1H). Based on these findings, CCF caused by a ruptured aneurysm of the PPTA trunk was diagnosed.

Under general anesthesia and using bilateral transfemoral vascular access, Optimo 8-Fr (Terumo, Tokyo, Japan) and CX 4-Fr (Terumo) guiding catheters were advanced into the left ICA and right VA, respectively. Simultaneous ICA and VA angiography demonstrated a well-defined road map for the PPTA aneurysm (Fig. 2A). A 7 × 7 mm Shouryu HR balloon occlusion catheter (Kaneka Medical Products, Osaka, Japan) was placed in conjunction with a Chi-

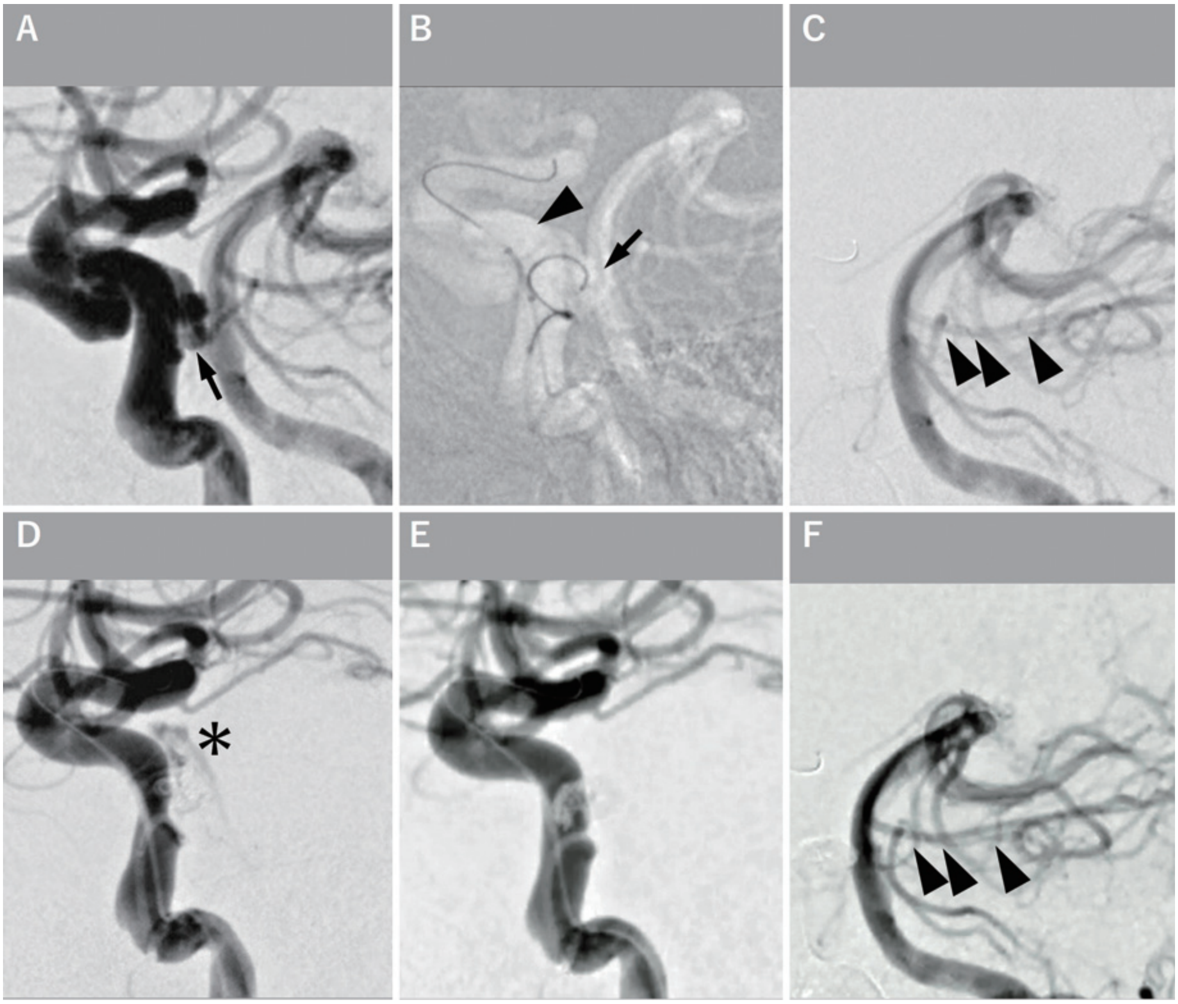

Fig. 2
A: Simultaneous internal carotid and vertebral angiography (lateral view as working angle) before treatment indicates an aneurysm of the persistent primitive trigeminal artery (PPTA) trunk (black arrow).
B: The road map view (lateral view as working angle) shows the microcatheter (black arrow) within the aneurysm and the balloon catheter (black arrowhead) in the C4 segment of the left internal carotid artery.
C: Right vertebral angiography (lateral view as working angle) during embolization indicates normal flow through the left anterior inferior cerebellar artery (black arrowheads).
D: Left internal carotid angiography (lateral view as working angle) during embolization indicates remnant shunt flow (asterisk).
E: Left internal carotid angiography (lateral view as working angle) after PPTA occlusion shows complete obliteration of the aneurysm and the PPTA trunk.
F: Right vertebral angiography (lateral view as working angle) during PPTA trunk embolization shows that the left anterior inferior cerebellar artery flow is patent (black arrowheads).

kai black 14 soft tip micro guidewire (Tokai Medical Products, Nagoya, Japan) in the C4 segment of the left ICA for flip-turn insertion of the catheter at the entrance of steep PPTA entrance (Fig. 2B). An Excelsior SL-10 microcatheter (Stryker Neurovascular, Fremont, CA, USA) was advanced into the aneurysm through the PPTA trunk using a Traxcess 14 microguidewire (MicroVention, Aliso Viejo, CA, USA) with a coaxial system consisting of a Tactics intermediate catheter (Tokai Medical Products) (Fig. 2B). Guiding the microcatheter to the distal side of the aneurysm (AICA side) was difficult because the PPTA was tortuous; therefore, we chose to perform intra-aneurysmal coil embolization. First, we formed a cage in the aneurysm using two detachable coils. A loop of the second coil deviated from the aneurysm into the CS, which indicated the fistulous point in the aneurysm. When the third coil was placed, shunt flow from the posterior circulation (from AICA via the PPTA) disappeared, while anterograde perfusion of the left AICA territory remained intact (Fig. 2C). ICA angiography showed flow from the left ICA to the shunt point (Fig.

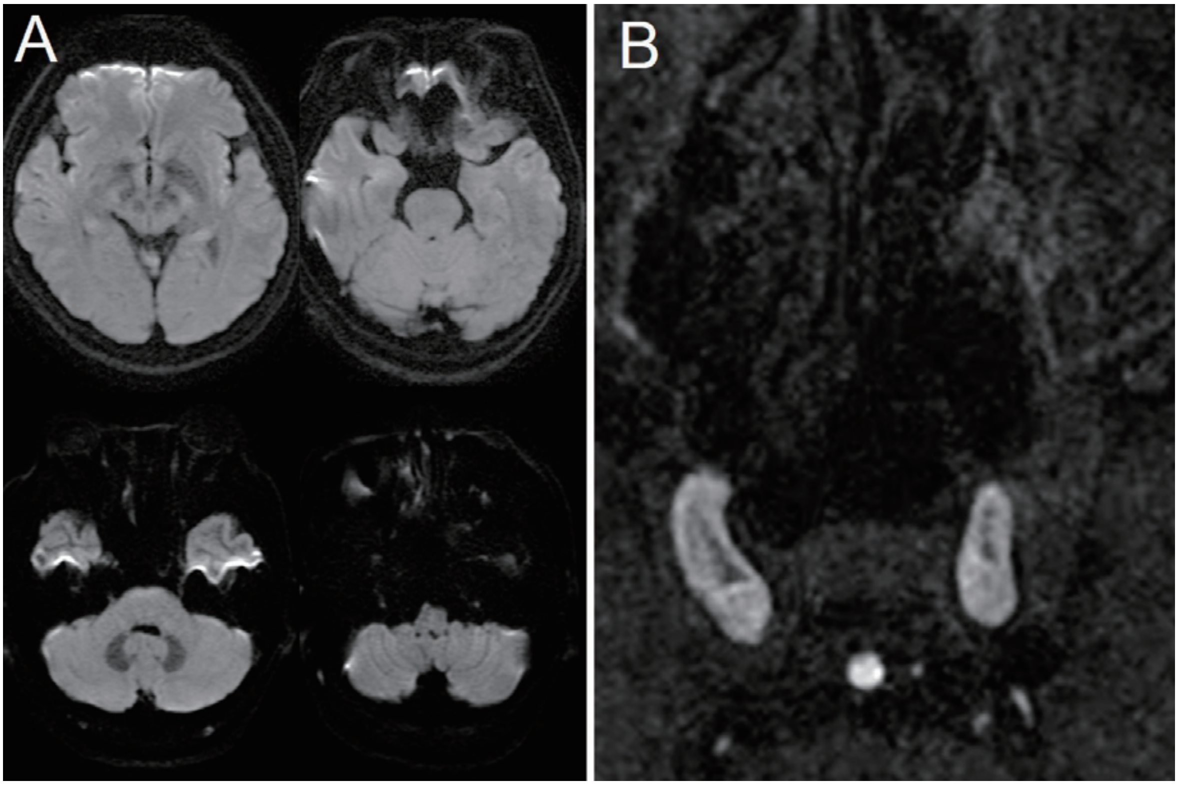

Fig. 3
A: Diffusion-weighted imaging after the procedure shows no ischemic lesions at the axial level of midbrain, pons, superior cerebellar peduncle, and medullar oblongata.
B: Time-of-flight magnetic resonance angiography at the axial level of the cavernous sinus shows disappearance of flow in the cavernous sinus and superior ophthalmic vein.

2D); therefore, internal trapping of the proximal side (ICA side) of the PPTA trunk was performed using detachable coils. Shunt flow disappeared after placement of 18 coils. At this point, the fistula and PPTA were completely occluded and shunt flow had stopped (Fig. 2E). VA angiography demonstrated normal perfusion of the posterior circulation (Fig. 2F). Immediately after the procedure, the patient's exophthalmos and hyperemia were alleviated. No new neurological symptoms developed and ischemic lesions were not observed on postoperative diffusion-weighted magnetic resonance imaging (Fig. 3A). The left CS, SOV, and IPS signal on time-of-flight magnetic resonance angiography also disappeared (Fig. 3B). There were no signs of recurrence on magnetic resonance imaging at the 3-year follow-up.

Discussion

The PPTA is one of the embryological anastomoses between the developing carotid artery and vertebrobasilar system that can persist into adulthood.[1] Saltzman classified the PPTA into three types based on angiographic findings in 1959.[7] The type 1 PPTA is commonly called a fetal persistent trigeminal artery since the BA is hypoplastic and the Pcom artery is absent; here, the PPTA is essential for providing blood flow to the posterior circulation. The type 2 PPTA is also called adult-type persistent trigeminal artery; here, the posterior cerebral artery and BA are principally supplied by the Pcom artery and VA, respectively. The type 3 PPTA is also known as PTAv; here, the artery arises from the proximal ICA and anastomoses with the superior, anterior inferior, or posterior inferior cerebellar artery.[8] Anastomosis to the AICA is most common (71.6%).[9] The PTAv results from inadequate fusion of the PPTA and the longitudinal neural artery. When present, the PTAv is the only source of cerebellar artery perfusion; however, in rare cases, the BA may connect with a cerebellar artery.[1] The PPTA in our patient was categorized as a PTAv that directly anastomosed with a left cerebellar cortical artery branch of AICA (itself a branch of the BA). Using a modern PPTA classification based on magnetic resonance angiography, it was categorized as type 5b connecting to the BA.[10]

To our knowledge, only nine cases of nontraumatic ruptured PTAv aneurysm have been reported, including ours.[9,11-15] Table 1 summarizes these cases. Four aneurysms were located at the ICA-PTAv junction and four on the parent artery. The mode of presentation was abducens

Table 1 Reported cases of nontraumatic persistent trigeminal artery variant aneurysmal rupture

Case No.	Author	Age and sex	Aneurysm site	Cerebellar artery	Symptom	Treatment
1	Hayashi et al., 1994	47, F	ICA-PPTA	PICA	SAH	Clipping
2	Hanabusa et al., 2000	71, F	PPTA trunk	AICA	SAH	Internal trapping
3	Nishio et al., 2001	69, F	ICA-PPTA	PICA	Abducens palsy	Embolization
4	Shin et al., 2005	40, M	ICA-PPTA	PICA	CF	TVE
5	Yang et al., 2010	48, M	PPTA trunk	AICA	SAH	Internal trapping
6	Yamamoto et al., 2011	82, F	PPTA trunk	AICA	SAH	Clipping
7	Matsumoto et al., 2011	61, F	PPTA trunk	PICA	Abducens palsy	Internal trapping
8	Takigawa et al., 2014	62, F	ICA-PPTA	SCA, AICA	(-)	Embolization
9	Present case	48, M	PPTA trunk	AICA	CF	Internal trapping

M, male; F, female; ICA, internal carotid artery; PPTA, persistent primitive trigeminal artery; PICA, posterior inferior cerebellar artery; SAH, subarachnoid hemorrhage; AICA, anterior inferior cerebellar artery; CF, cavernous fistula; TVE, transvenous embolization; SCA, superior cerebellar artery

nerve palsy in two patients, subarachnoid hemorrhage in four, cavernous fistula in two, and no symptoms in one. Endovascular treatment was performed in six—aneurysm embolization in two and internal trapping in four. There were no cerebral infarction complications in any of the six.

In our patient, the CCF was a high-flow shunt and the shunt point was difficult to identify. Imaging during the balloon occlusion test helped to identify the shunt position and the anastomosis to the BA, which is rare. Simultaneous angiography of the ICA and the VA was also crucial for creating a well-defined road map for transarterial embolization.

We performed internal trapping of the PTAv trunk. Although transvenous embolization is generally considered a superior method for achieving complete shunt occlusion, it carries a risk of injury to the cranial nerves traversing the CS. In addition, particularly in cases of aneurysm rupture, transvenous embolization may not be sufficient, which would result in persistence of symptoms.

In our patient, the shunt point was a fusiform-like aneurysm and PTAv trapping was unavoidable to obtain a sufficient embolizing effect. However, trapping cut off the PTAv blood flow; therefore, it was necessary to evaluate the risk of ischemia in the PTAv territory, namely, the cerebellar cortical artery region and areas supplied by PTAv penetrating branches. In addition, his PTAv connected to the BA, which is rare. If blood flow from the anastomotic BA is sufficient, then arterial flow to the cerebellar cortex can be maintained. Vertebral angiography during the ICA balloon occlusion test demonstrated an anastomotic BA that was obscured by anterograde flow from the PTAv.

The existence of “invisible” penetrating branches from the main trunk that are not shown owing to the adjacent high-flow shunt can pose a problem. Although an accurate evaluation is difficult, we believe that the risk of infarction in these branches may not necessarily be very high because no ischemic symptoms were shown by preliminary balloon occlusion test of the proximal ICA, which cut off dominant anterograde blood flow to the PTAv although retrograde alternative flow was not blocked. In addition, no ischemic complications have been previously reported by internal trapping of the PTAv. Performing an occlusion test at the level of the PTAv bifurcation may allow further evaluation of ischemic tolerance, but blood flow to the PTAv from the posterior circulation remains, so it may still be an incomplete occlusion test. Short segment embolization may be useful if the catheter can be guided to the BA side of the aneurysm to reduce the risk of perforator infarction.

We were able to achieve occlusion of the shunt point between the PPTA and CS via internal trapping. Two catheters were used, one in the ICA-PPTA to perform internal trapping and perform anterior angiography and another in the BA for performing posterior angiography. This aids in the identification of the shunt point during vessel occlusion from both the anterior and posterior sides and prevents ischemia in the AICA territory. Our technique should be considered by fellow neurointerventionalists.

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Ethics Approval

The Ethics Committee for Human Research at our institution approved the study (IRB no. R2023-076). The study was conducted in accordance with the Declaration of Helsinki.

Conflicts of Interest Disclosure

All authors declare no conflicts of interest.

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