

Carotid-Cavernous Sinus Fistula caused by Persistent Primitive Trigeminal Artery Aneurysm rupture: A case report

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Carotid-Cavernous sinus fistula (CCF) caused by a ruptured aneurysm of persistent primitive trigeminal artery (PPTA) is exceptionally rare. Herein, we reported the case of a 64-year-old female who presented with headache, pulsatile tinnitus, and diplopia. Angiography showed a PPTA and CCF. Three-dimensional digital subtraction angiograms revealed a direct shunt between the PPTA aneurysm and the left cavernous sinus. The trigeminal carotid fistula was successfully treated with detachable coils and Onyx, and the PPTA was preserved. Hence, coil and Onyx occlusion of the cavernous sinus is a safe and effective treatment for trigeminal carotid fistula.

Key Words: Carotid-cavernous sinus fistula—persistent primitive trigeminal artery—aneurysm—endovascular therapy

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Introduction

Persistent primitive trigeminal artery (PPTA) is a common anatomical variation of carotid-basilar artery during adulthood, with a reported incidence of .1%-.6% based on conventional angiography. The PPTA was first identified during autopsy by Quain in 1844, and also in an angiography study by Sutton in 1950.¹ Primitive trigeminal artery forms a link between the basilar artery and internal carotid artery during embryogenesis, which usually arises from

the cavernous sinus internal carotid artery, followed by anastomosis to the basilar artery between the superior and anterior inferior cerebellar arteries. A ruptured aneurysm of the PPTA results in carotid-cavernous sinus fistula (CCF), which are exceptionally rare. Herein, we presented a case of CCF caused by a ruptured PPTA aneurysm, which was successfully treated by detachable coil and Onyx. The Ethics Committee approved the case report, which was in compliance with the Helsinki Declaration.

Case Report

A 64-year-old female presented with a 6-month history of headache and pulsatile tinnitus in her auris sinister, without any history of head trauma. She also suffered from diplopia and exophthalmos in her left eye for 1 month. Physical examination on admission showed left abducens nerve palsy, and a periorbital pulsatile bruit. Noncontrast computed tomography revealed an enlarged cavernous sinus and superior ophthalmic vein (Fig 1). Left internal carotid digital subtraction angiography (DSA) revealed a carotid cavernous sinus fistula, with major drainage from superior ophthalmic vein to anterior facial vein and then onto external jugular vein. Lateral view of left internal carotid DSA showed a connection of internal carotid and basilar artery with PPTA (Fig 2A), and an aneurysm of PPTA projecting into the left cavernous sinus (Fig 2B). Three-dimensional

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Received May 22, 2019; revision received June 26, 2019; accepted July 21, 2019.

Financial Disclosure: This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

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1052-3057/\$ - see front matter

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<https://doi.org/10.1016/j.jstrokecerebrovasdis.2019.104306>



Figure 1. Arrowheads show enlarged cavernous sinus and superior ophthalmic vein.

DSA revealed a ruptured aneurysm at the origin of PPTA trunk and blood flow into the cavernous sinus (Fig 2C). Left vertebral angiography showed retrograde blood flow filling of the PPTA and CCF after compressing the common carotid artery (Fig 2D). The clinical symptoms of the patient were mainly due to high pressure in the cavernous sinus.

The endovascular treatment plan involved passing a microcatheter through PPTA aneurysm crevasse into cavernous sinus, followed by embolization of CCF with detachable coils and Onyx-18 liquid embolic system (Covidien, Irvine, CA). Under general anesthesia, 3000 units of heparin were intravenously administered to the patient. A 6 Fr Envoy guiding catheter (Cordis, Miami Lake, FL) was introduced through a femoral sheath into the petrous portion of left internal carotid artery over a .035-inch steerable guide wire (Terumo Company, Tokyo), using road mapping technique. During endovascular treatment, it is difficult to superselect the fistula through aneurysm sac. Echelon-10 (Medtronic, Irvine, CA) and Excelsior SL-10 microcatheter (Boston Scientific, Fremont, CA) guided by Traxcess-14 (MicroVention/Terumo, Tustin, CA) and PT Graphix wire had previously failed. So, we inserted a flexible Headway

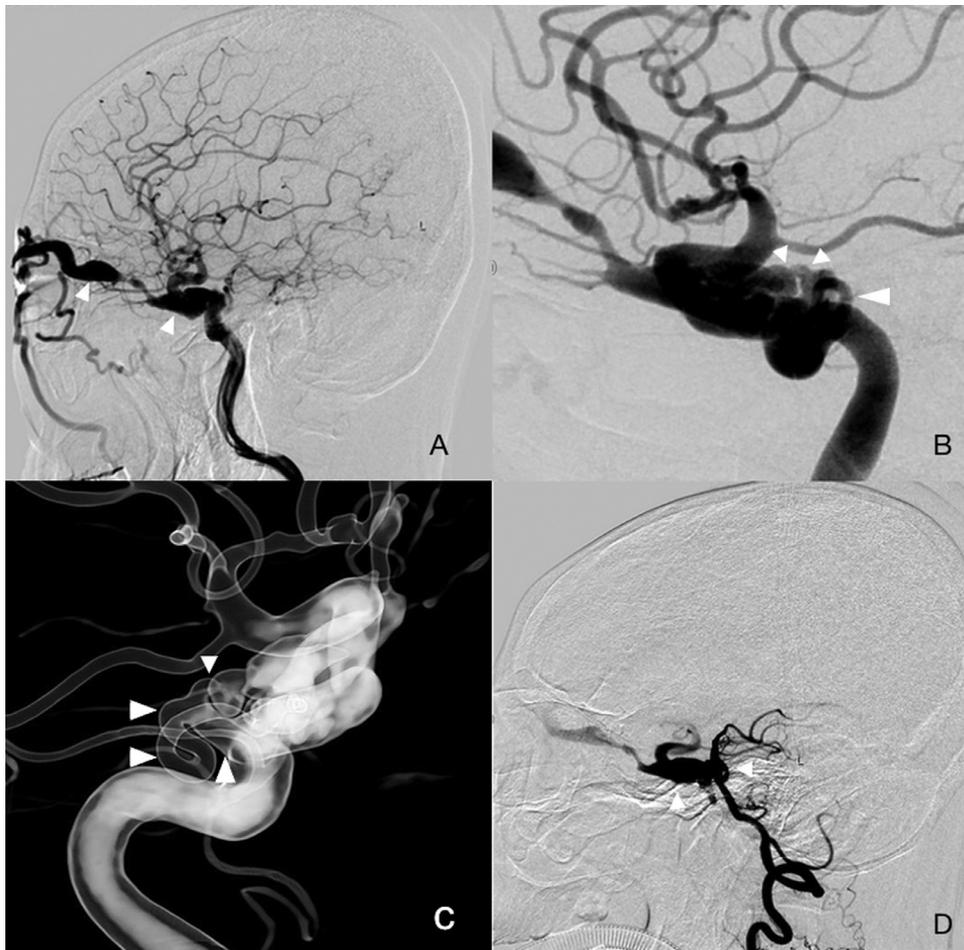


Figure 2. (A) Lateral view of left internal carotid DSA showed a CCF. Superior ophthalmic vein (arrowheads); (B) CCF caused by PPTA (long white arrow) aneurysm (white arrow) rupture; (C) Three-dimensional DSA revealed PPTA (long white arrow) trunk aneurysm (white arrow); (D) Compressed the common carotid artery, left vertebral angiography, lateral view, showed PPTA and CCF.

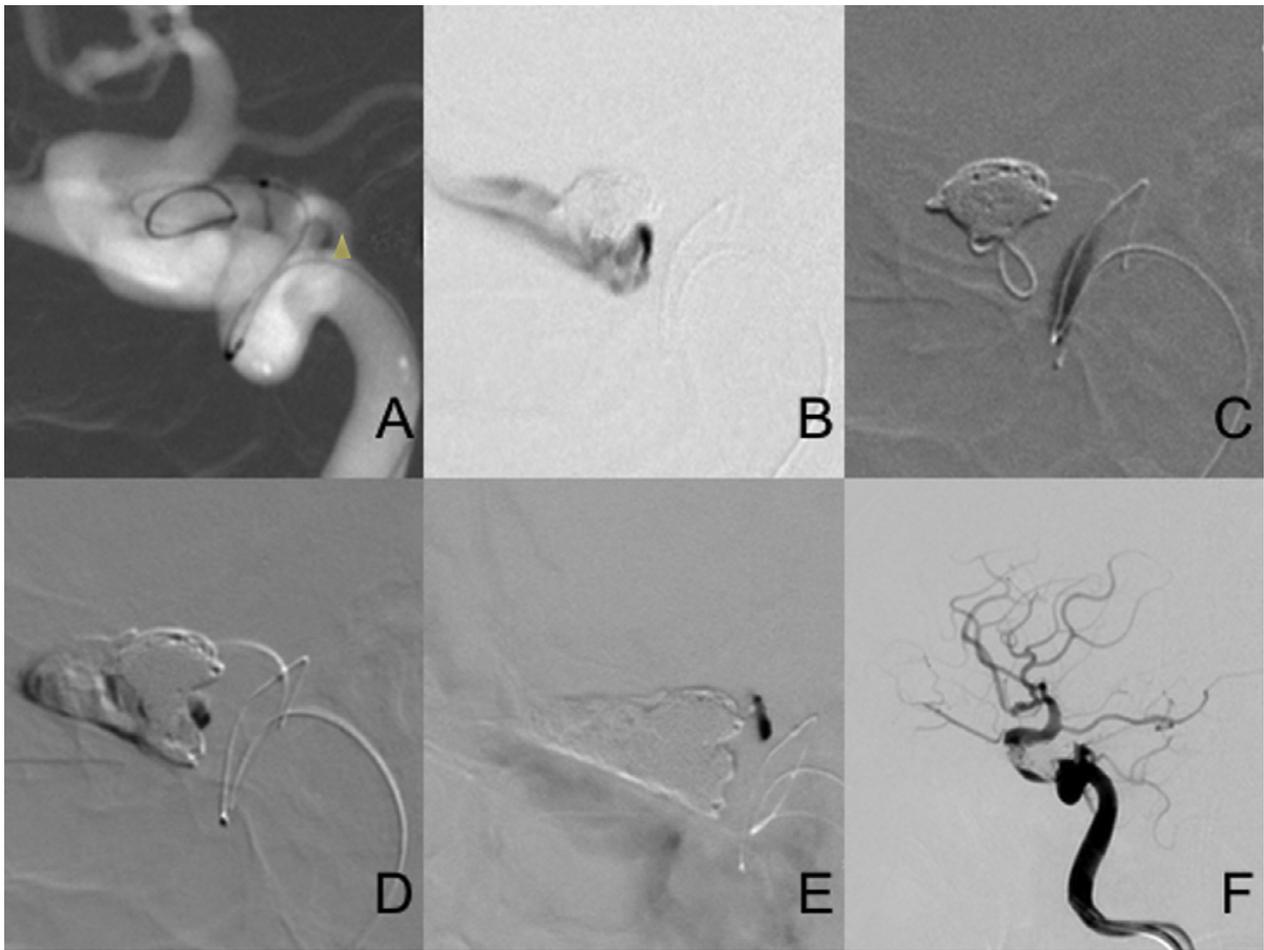


Figure 3. Digital subtraction angiograms performed during embolization. (A) insertions of the first coil, The arrow points to PPTA; (B) the second coil; (C) Filling HyperGlide balloon; (D, E) injection of Onyx. When part of Onyx enters PPTA aneurysm, stop injection to prevent Onyx from entering basilar artery; (F) the CCF completely disappeared with preservation of the persistent primitive trigeminal artery.

Duo microcatheter into the sinus through PPTA aneurysm crevasse, guided by a reliable stability PT Graphix wire. Then, the cavernous sinus was carefully embolized by two AXIUM (Medtronic, Irvine, CA) detachable coils. HyperGlide balloon 4 × 20-mm (Medtronic, Irvine, CA) occlusion protected the left internal carotid artery, 2 mL Onyx-18 liquid embolic system embolized the CCF and part of the PPTA aneurysm. Finally, the CCF completely disappeared, while the PPTA was preserved (Fig 3).

The patient had favorable outcome. The pulsatile tinnitus and exophthalmos disappeared 1 day after the procedure, while the left abducens nerve paralysis recovered within 3 months.

Discussion

The presence of PPTA in adulthood is due to angiodysplasia in cerebrovascular development, and may be associated with vascular anomalies such as aneurysms.² Kai et al found that about 15% of PPTA are combined with aneurysm.³ The PPTA aneurysm is thought to be due to the presence of structural defects in the vascular walls or

hemodynamic stress on the PPTA, but there is no histological evidence to support this hypothesis.⁴ PPTA aneurysm rupture may lead to subarachnoid hemorrhage or CCF.⁵ However, CCF rupture of an aneurysm at origin or proximal trunk of the PPTA is exceptionally rare. A literature review in PubMed revealed only 9 cases of PPTA rupture aneurysm that led to CCF, including the present case (Table 1). The age of the patients ranged from 42 to 64 years. The main symptoms included headache, exophthalmos, chemosis, ophthalmoplegia, diplopia, and pulsatile bruit. The locations of the fistula were at the junction of internal carotid artery and PPTA in 5 cases, and at the PPTA trunk in 7 cases. Yoshida et al⁶ presented a case that clearly showed the PPTA trunk aneurysm by 3-dimensional digital subtraction angiograms (3D-DSA). Ruptured PPTA aneurysm can be rarely detected by cerebral angiography because of the high flow of fistula that causes the contrast agent to quickly fill the cavernous sinus, and the destruction of the aneurysmal sac.⁷ As in the present case, the residual aneurysm sac can be seen by 3D-DSA. Therefore, we recommend 3D-DSA to examine a CCF caused by PPTA aneurysm in order to determine the

Table 1. Summary of previously reported cases of CCF caused by rupture aneurysm of PPTA

Case	Author (Year)	Age, sex	Symptoms	Location	Treatment	Outcomes
1	Enomoto et al (1977) ⁸	42, F	Ophthalmic pain, proptosis, diplopia	ICA-PPTA (right)	ICA ligation	Cure, mild ophthalmoplegia
2*	Charli et al (1982) ⁶	53, F	Ocular disorder	PPTA trunk (left)	Balloon	Cure
3*	Kodama et al (1984) ⁹	53, F	-	ICA-PPTA (right)	Clipping	Total ophthalmoplegia
4	Guglielmelli et al. (1990) ¹¹	57, M	Diplopia, bruit, ophthalmoplegia	ICA-PPTA (left)	Balloon	Cure
5	Xin-Yak Qian et al (2009) ¹²	62, F	Eye pain, chemosis, ophthalmoplegia, diplopia, bruit	ICA-PPTA (right)	Coil (transvenous)	Cure
6	Liu et al (2009) ¹³	55, F	Diplopia, headache, pulsatile bruit	PPTA trunk (left)	Coil (transarterial)	Cure
7	Kim et al (2010) ¹⁴	42, F	Diplopia, headache, exophthalmos	PPTA trunk (left)	Coil (transarterial)	Cure
8	Yoshida et al (2011) ⁶	60, F	Bruit, ophthalmoplegia	PPTA trunk (left)	Coil (intra-an)	Cure
9	Present case	64, F	Diplopia, headache, pulsatile bruit, exophthalmos	PPTA trunk (left)	Coil & Onyx (transarterial)	Cure

Abbreviations: F, female; M, male; ICA, internal carotid artery; PPTA; persistent primitive trigeminal artery.

*The original full text was not found.

relationship of vessel and the presence of aneurysms before planning the treatment strategy. The treatment methods included ligation or direct surgery in 2 cases,^{8,9} detachable balloons in 4 cases,^{5,6,10,11} and coil embolization in 4 cases.^{6,12-14} The present case was treated with transarterial coiling and Onyx-18 liquid embolic system. It is necessary to consider whether the PPTA can be occluded when formulating therapeutic strategies. Saltzman type 1 is when the PPTA supplies the posterior cerebral and superior cerebellar territories, which should not be sacrificed. The present case was Saltzman type 2 wherein the PPTA anastomoses with basilar artery, but mainly supplies the bilateral SCAs.⁵ For type 2, the PPTA can be embolized.

The prevalent treatment for CCF is detachable balloon embolization. Transarterial approach is recommended for balloon and coil embolizations. Transvenous cavernous sinus packing with detachable coils may require a large volume of coils, but should be considered in case of difficulty with a transarterial approach.¹⁵ Given the PPTA aneurysm and small size of the fistula in this case, detachable balloon embolization or intra-aneurysm coiling were difficult. Therefore, the treatment strategy adopted for this patient was transarterial sinus packing with coils, followed by injection of Onyx with balloon protection, and subsequent complete closing of the fistula with preservation of the internal carotid artery and PPTA. While injecting the Onyx, we only preserved the internal carotid artery with balloon assistance. However, it would be safer if a balloon is used to simultaneously protect the basilar artery.

Declaration of Competing Interest

All authors declare no competing interests.

Ethical Approval

The Ethics Committee of Baotou Central Hospital approved the research protocol in compliance with the Helsinki Declaration.

Informed Consent

Informed consent was obtained from this patient included in the study.

References

- O'uchi E, O'uchi T. Persistent primitive trigeminal arteries (PTA) and its variant (PTAV): analysis of 103 cases detected in 16,415 cases of MRA over 3 years. *Neuroradiology* 2010;52:1111-1119. <https://doi.org/10.1007/s00234-010-0669-6>.
- Murai Y, Kobayashi S, Tateyama K, et al. Persistent primitive trigeminal artery aneurysm associated with cerebellar hemangioblastoma. Case report. *Neurol Med Chir* 2006;46:143-146.
- Kai Y, Ohmori Y, Watanabe M, et al. Coil embolization of an aneurysm located at the trunk of the persistent primitive trigeminal artery. *Neurol Med Chir* 2011;51:361-364.
- Takase T, Tanabe H, Kondo A, et al. Surgically treated aneurysm of the trunk of the persistent primitive trigeminal artery—case report. *Neurol Med Chir* 2004;44:420-423.
- McKenzie JD, Dean BL, Flom RA. Trigeminal-cavernous fistula: Saltzman anatomy revisited. *Am J Neuroradiol* 1996;17:280-282.
- Yoshida M, Ezura M, Mino M. Carotid-cavernous fistula caused by rupture of persistent primitive trigeminal artery trunk aneurysm—case report. *Neurol Med Chir* 2011;51:507-511.
- Cheng WC, Wang AD. Carotid-cavernous sinus fistula associated with a primitive trigeminal artery. *Neurosurgery* 1990;27:802-805.
- Enomoto T, Sato A, Maki Y. Carotid-cavernous sinus fistula caused by rupture of a primitive trigeminal artery

- aneurysm. Case report. *J Neurosurg* 1977;46:373-376. <https://doi.org/10.3171/jns.1977.46.3.0373>.
9. Oka Y, Sadamoto K, Tagawa M, Fujita M, et al. Transvenous embolization of carotid-cavernous sinus fistula associated with a primitive trigeminal artery—case report. *Neurol Med Chir* 2000;40:61-64.
 10. Flandroy P, Lacour P, Marsault C, et al. The intravascular treatment of a cavernous fistula caused by rupture of a traumatic carotid trigeminal aneurysm. *Neuroradiology* 1987;29:308-311.
 11. Guglielmi G, Vinuela F, Dion J, et al. Persistent primitive trigeminal artery-cavernous sinus fistulas: report of two cases. *Neurosurgery* 1990;27:805-808. discussion 808–9.
 12. Qian CX-Y, Ares C, Codere F, et al. Rupture of an aneurysm of the persistent trigeminal artery presenting as a carotid-cavernous sinus fistula. *Orbit* 2009;28:275-280. <https://doi.org/10.3109/01676830903190222>.
 13. Liu L, He H, Li Y, et al. Rupture of persistent primitive trigeminal artery aneurysm associated with a cavernous sinus fistula. A case report and review of the literature. *Neuroradiol J* 2009;22:471-475. <https://doi.org/10.1177/197140090902200419>.
 14. Kim BM, Kim DI, Kwon TH. Persistent Trigeminal artery with a cerebellar branch and trigeminal-cavernous fistula from ruptured aneurysm: transarterial coil embolization. *Neurointervention* 2010;5:32. <https://doi.org/10.5469/neuroint.2010.5.1.32>.
 15. Kobayashi N, Miyachi S, Negoro M, et al. Endovascular treatment strategy for direct carotid-cavernous fistulas resulting from rupture of intracavernous carotid aneurysms. *Am J Neuroradiol* 2003;24:1789-1796.