

## Case Report

# Direct Carotid Cavernous Fistula of an Adult-Type Persistent Primitive Trigeminal Artery with Multiple Vascular Variations

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We report a case of spontaneous right carotid-cavernous fistula (CCF) in a proximal segment of persistent primitive trigeminal artery (PPTA) and combined vascular anomalies such as left duplicated hypoplastic proximal posterior cerebral arteries and a variation of anterior choroidal artery supplying temporal and occipital lobe. A 45-year-old male presented with progressive right exophthalmos, diplopia, and ocular pain. With manual compression of the internal carotid artery, a cerebral angiography revealed a right CCF from a PPTA. Treatment involved the placement of detachable non-fibered and fibered coils, and use of a hyperglide balloon to protect against coil herniation into the internal carotid artery. A final angiograph revealed complete occlusion of PPTA resulted in no contrast filling of CCF.

**Key Words :** Persistent primitive trigeminal artery · Carotid cavernous fistula · Duplicated posterior cerebral artery.

## INTRODUCTION

The persistent primitive trigeminal artery (PPTA) is the most common embryological carotid-basilar anastomosis to persist into adulthood, reported to be incidentally found in 0.1-0.6% of all cerebral angiograms<sup>3</sup>.

Carotid-cavernous fistula (CCF) from PPTA to cavernous sinus and its endovascular treatment using detachable or fibered coils have been reported<sup>1,2,6,8,10,11</sup>. However, to my knowledge, detailed description of ruptured point and combined vascular variations in trigeminal fistula has not been published in the literature. We describe a patient with spontaneous CCF of an adult-type PPTA, which had combined multiple vascular variations, treated with coil embolization via arterial route.

## CASE REPORT

A 46-year-old male with no medical illness and no trauma

history presented with complaints of right exophthalmos, right ocular pain, tinnitus, and diplopia. These symptoms persisted and aggravated gradually during 2 months. His visual acuity was not impaired. Lateral view of right internal carotid artery (ICA) angiogram showed a typical appearance of CCF with its major venous drainage into ipsilateral superior ophthalmic vein and inferior petrosal sinus and variation of anterior choroidal artery supplying to occipital and temporal lobe (Fig. 1A). Right carotid angiography revealed a fistulous tract originated from the posterior vertical cavernous segment of right ICA under manual compression of cervical carotid artery without visualization of PPTA (Fig. 1B). Towne's and lateral view of right vertebral angiogram with compression of carotid artery revealed contrast filling of PPTA directly to cavernous sinus and superior ophthalmic vein and sylvian cortical vein, hypoplastic duplicated proximal posterior cerebral arteries (PCAs), and duplicated anterior inferior cerebellar artery (AICA) (Fig. 1C). Magnified Selective microangiogram showed fistulous hole not in junction between cavernous ICA and PPTA origin but in proximal PPTA inside cavernous sinus (Fig. 1D). Endovascular treatment was performed immediately after full angiographic evaluation of CCF. After placement of 6F introducer sheath in the bilateral femoral arteries, 6F guiding catheter (Envoy; Cordis, Miami, FL, Guider softtip; Boston Scientific, Fremont, CA, USA) was positioned to the cervical segment of right ICA and

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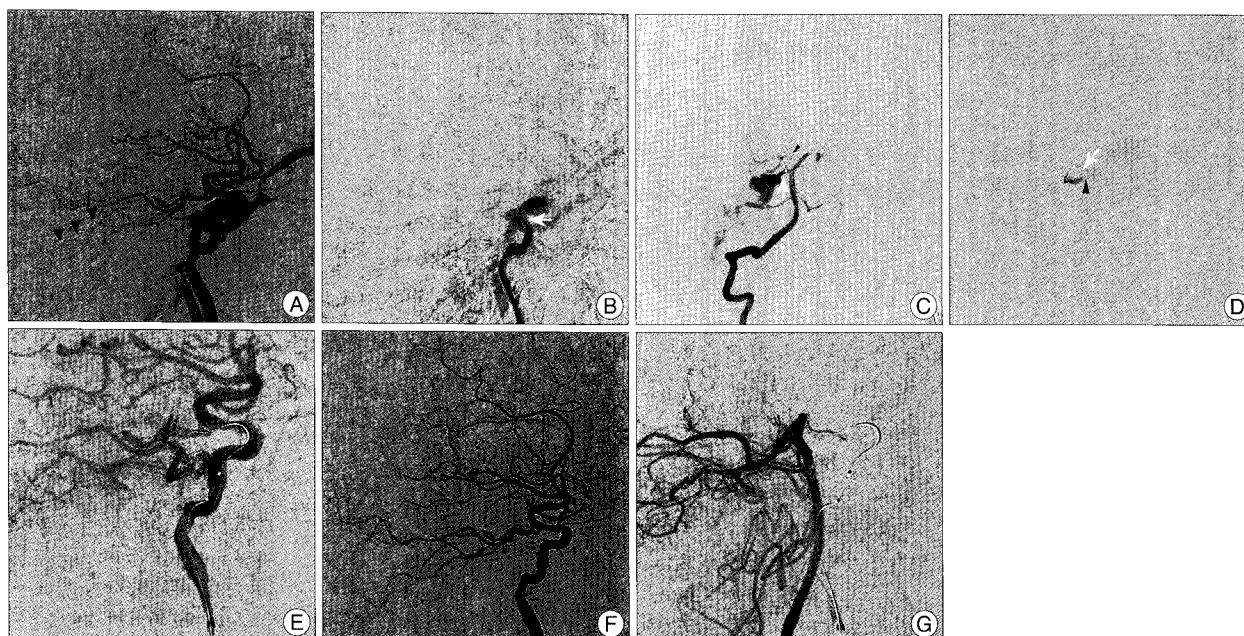
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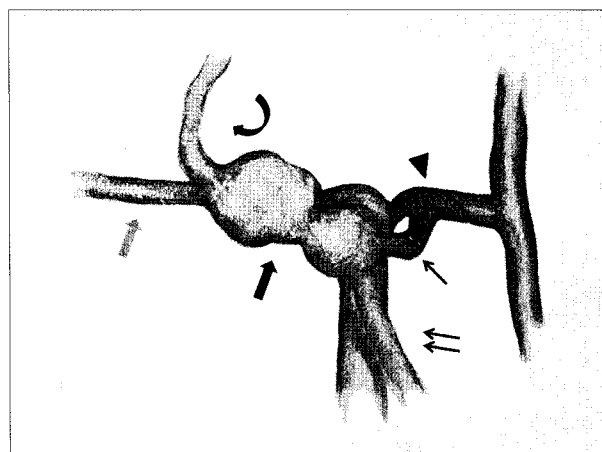
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**Fig. 1.** A : Initial lateral view of right internal carotid artery (ICA) angiogram showing variation of anterior choroidal artery supplying to temporal and occipital lobe (multiple arrow heads) and the typical appearance of a carotid-cavernous fistula (CCF) with main venous drainage through cavernous sinus (CS) and petrosal sinus (PS). B : Lateral view under manual compression of carotid artery revealing a single fistulous tract (arrow) between right ICA and CS. C : Anteroposterior view of right vertebral angiogram under manual compression of cervical carotid artery demonstrating duplicated hypoplastic proximal posterior cerebral arteries (arrow head), left proximal segment of superior cerebellar artery (white arrow head) without contrast filling of distal portion, and contrast filling of CCF and inferior PS through persistent primitive trigeminal artery (PPTA) (black arrow). D : Selective microangiogram revealed that fistulous hole was not junction between the origin of PPTA and cavernous ICA (arrow) but proximal segment of PPTA (arrow head). E : Four detachable bare platinum coils were packed into the fistulous tract flow through the fistulous tract was occluded and more clear opacification of the proximal PPTA appeared on control angiogram. F and G : Lateral ICA angiogram and vertebral angiogram immediately after embolization showed complete occlusion of the CCF and the PPTA.

V2 segment of right vertebral artery. A 4×10 mm Hyperglide balloon (ev3 Neurovascular, Irvine, CA, USA) with silver speed microwire (ev3 Neurovascular, Irvine, CA, USA) was placed to cavernous segment of right ICA covering fistulous tract. The origin of PPTA was selected with by using microcatheter (Excelsior 10; Boston Scientific) and 0.010-inch guidewire (Agility; Cordis) by using the coaxial technique through right ICA. Further advance of microcatheter into cavernous sinus was difficult due to vascular tortuosity. Occlusion of proximal PPTA including fistulous hole using coils via transarterial route was considered. After embolization of PPTA with fistulous point by using five detachable bare platinum coils under balloon protection of cavernous ICA, there was no flow through the fistulous tract and prominent appearance of proximal PPTA (Fig. 1E). Further embolization using two detachable fibered coil and one bare platinum coil (GDC; Boston Scientific, Watertown, MA, USA) was performed in the fistulous tract. A postembolization of right internal carotid angiogram and vertebral angiogram showed complete occlusion of PPTA and fistulous point resulted in non-visualization of CCF (Fig. 1F, G). The patient recovered from right ocular pain and diplopia immediately after the treatment. At clinical follow up one month after embolization, his exophthalmos and tinnitus disappeared completely. Brain magnetic resonance angiogram confirmed complete occlusion of the fistula and no evidence of brain ischemia.



**Fig. 2.** Schematic illustration demonstrating the relationships among persistent primitive trigeminal artery (black arrowhead) with fistulous hole (single arrow), cavernous sinus (black solid arrow), superior ophthalmic vein (grey solid arrow), cortical drainage vein (curved arrow), and inferior petrosal sinus (double arrow).

## DISCUSSION

The most frequent cause of direct CCF are trauma, which accounts for 70 to 90% of cases. Spontaneous direct CCFs are often associated with underlying collagen deficiencies, such as Ehlers-Danlos syndrome<sup>12</sup>. However, spontaneous direct CCFs

are usually caused by rupture of an cavernous aneurysm, and incidence of these was reported to be about 20%<sup>9)</sup>. Ruptured aneurysm is often very difficult to detect in the CCF by cerebral angiography, because of the high-flow fistula and the destruction of the aneurysm sac<sup>10)</sup>. In the present case, angiography did not identify the aneurysmal dilatation as well. Selective microangiogram revealed a precise point of fistulous tract not in junction between PPTA origin and cavernous ICA but in the proximal PPTA inside cavernous sinus. Vascular anomaly of unknown etiology in the proximal PPTA inside cavernous sinus might probably be a cause of spontaneous CCF in our case.

Classification of persistent primitive trigeminal artery<sup>4)</sup> was as follow : 1) fetal type in which the posterior circulation is dependent on the anastomosis via PPTA, and 2) adult type where the posterior circulation is independent of the anastomosis. The risk of posterior circulation infarction is dependent on the type of the PPTA and the native diameter of basilar artery. With a large basilar artery, the risk of posterior circulation infarction is relatively small in the endovascular occlusion of adult type of PPTA.

Treatment of CCF is to fill in fistulous tract completely and maintain the patency of the ICA. Selective transarterial balloon embolization is the preferred treatment for direct CCF. Coil embolization can be performed if the lesion is not amendable to balloon occlusion. However, detachable balloon has not been used for several years in our country. Therefore, coil embolization via arterial approach was planned for our patient initially. Awareness of the correct fistular hole and the type of PPTA enabled us to perform coil embolization of proximal PPTA including fistulous tract using only arterial approach. In the present case, occlusion of proximal PPTA including ruptured point may be an useful technique in the treatment of PPTA fistula. The most important risk of coil embolization is unwanted coil migration into the ICA. Techniques to prevent the coil dislocation include the placement of a nondetachable balloon or self-expandable stent in the ICA across the origin of the fistula during coil packing<sup>5,7)</sup>. In the present case, balloon protection rather than stent from unwanted coil loop herniation into the ICA was considered because premedication such as aspirin and clopidogrel is required if a stent is used.

True duplicated hypoplastic P1 segment is a unique variation of PCA which is caused by abnormality in fusion process of basilar artery. This may be well balanced with incomplete regression of PPTA, prominent posterior communicating artery, and anterior choroidal artery variant supplying to temporal and occipital lobe in the aspect of vascular maturization and hemo-

dynamic equilibrium.

## CONCLUSION

The present report describes a rare case of a spontaneous CCF caused by an unknown vascular anomaly in the proximal PPTA with combined vascular variations. Transarterial embolization using a balloon to protect against unwanted coil herniation into the ICA appears to be a successful treatment strategy in such cases.

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