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.
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 [12]. However, spontaneous direct CCFs...
are usually caused by rupture of an cavernous aneurysm, and incidence of these was reported to be about 20%. Ruptured aneurysm is very difficult to detect in the CCF by cerebral angiography, because of the high-flow fistula and the destruction of the aneurysm sac. 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 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. 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 maturatization and hemodynamic 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.

**References**

1. Bernstein K, Teitelbaum GP, Herman B, Giannotta SL: Coil embolization of a trigeminal-cavernous fistula. AJNR Am J Neuroradiol 19: 1953-1954, 1998
2. Chan YL, Shing KK, Wong KC, Poon WS: Transvenous embolisation of a carotid-trigeminal cavernous fistula. Hong Kong Med J 12: 310-312, 2006
3. Cheng WC, Wang AD: Carotid-cavernous sinus fistula associated with a primitive trigeminal artery. Neurosurgery 27: 802-805, 1990
4. Cook BE Jr, Leavitt JA, Dolan JW, Nichols DA: Carotid cavernous fistula associated with persistent primitive trigeminal artery. J Neuroophthalmol 20: 264-265, 2000
5. Eddleman CS, Surdell D, Miller J, Shaibani A, Bendok BR: Endovascular management of a ruptured cavernous carotid artery aneurysm associated with a carotid cavernous fistula with an intracranial self-expanding microstent and hydrogel-coated coil embolization: case report and review of the literature. Surg Neurol 68: 562-567; discussion 567, 2007
6. Geibprasert S, Jararakmongk P, Krings T, Pongpech S: Trigeminal fistula treated by combined transvenous and transarterial embolisation. Acta Neurochir (Wien) 150: 583-588, 2008
7. Halbach VV, Higashida RT, Hieshima GB, Hardin CW, Yang PJ: Transvenous embolization of direct carotid cavernous fistulas. AJNR Am J Neuroradiol 9: 741-747, 1988
8. Hurst RW, Howard RS, Zager E: Carotid cavernous fistula associated with persistent trigeminal artery: endovascular treatment using coil embolization. Skull Base Surg 8: 225-228, 1998
9. Lewis AL, Tomesick TA, Tew JM Jr: Management of 100 consecutive direct carotid-cavernous fistulas: results of treatment with detachable balloons. Neurosurgery 36: 239-244; discussion 244-245, 1995
10. Oka Y, Sadamoto K, Tagawa M, Kuman Y, Sakai S, Fujita M: Transvenous embolization of carotid-cavernous sinus fistula associated with a primitive trigeminal artery—case report. Neurol Med Chir (Tokyo) 40: 61-64, 2000
11. Tokunaga K, Sugii K, Kameda M, Sakai K, Terasaka K, Higashi T, et al.: Persistent primitive trigeminal artery–cavernous sinus fistula with intracerebral hemorrhage: endovascular treatment using detachable coils in a transarterial double-cather technique. Case report and review of the literature. J Neurosurg 101: 697-699, 2004
12. Vinuela F, Fox AJ, Debrun GM, Peerless SJ, Drake CG: Spontaneous carotid-cavernous fistulas: clinical, radiological, and therapeutic considerations. Experience with 20 cases. J Neurosurg 60: 976-984, 1984