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Case Report

Infratentorial arteriovenous malformation associated with persistent primitive hypoglossal artery

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Abstract

Background: We report a case of infratentorial arteriovenous malformation (AVM) associated with persistent primitive hypoglossal artery (PPHA). To our knowledge, this is the second reported case of these combined anomalies in the English literature. We discuss the embryological relationship between these two congenital vascular anomalies.

Case Description: An 18-year-old girl, who suddenly developed severe headache and vomiting followed by loss of consciousness, was admitted to our hospital. A computed tomography scan showed intracerebellar hemorrhage with obstructive hydrocephalus. Digital subtraction angiography revealed an AVM in the left cerebellar hemisphere and an ipsilateral PPHA. After the intracranial pressure was stabilized, the AVM was surgically removed. AVMs develop during the 4th to 8th week of embryonic life. In contrast, carotid-basilar anastomoses (CBAs) including primitive hypoglossal arteries appear and close spontaneously by the 6th week of embryonic life. Thus, AVMs precede CBAs, and a large amount of blood flows into the adjoining AVM via ipsilateral CBAs. As a result, spontaneous closure of a CBA may be disturbed.

Conclusion: We speculate that coexistence of infratentorial AVMs and ipsilateral CBAs is not incidental but inevitable.

Key Words: Arteriovenous malformation, carotid-basilar anastomosis, posterior fossa, persistent primitive hypoglossal artery

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INTRODUCTION

Arteriovenous malformations (AVMs) are congenital vascular anomalies that arise as a result of abnormal blood vessel development during the early embryonic period.[17] Of all AVMs, infratentorial AVMs account for 6-8%.[8,15]

Persistent carotid-basilar anastomosis (CBA) is also a congenital vascular anomaly. It includes persistent primitive trigeminal artery (PPTA), persistent primitive hypoglossal artery (PPHA), persistent primitive otic artery, and persistent primitive proatlantal artery (type 1 and type 2).[23] The angiographic incidence of PPHA and PPTA are 0.05% and 0.20%, respectively.[19]

To the best of our knowledge, the incidence of infratentorial AVM with CBA is 2-3 times higher than that of common infratentorial AVM.[1-16,18,20-22,25-32] In addition, all reported infratentorial AVMs were

located in the cerebellar hemisphere ipsilateral to the CBA.[6,21,23,31] We herein report a case of infratentorial AVM associated with PPHA and discuss the embryologic relationship between these congenital vascular anomalies.

Case descriptions

An 18-year-old girl presented with a ruptured infratentorial AVM. The patient's Glasgow Coma Scale score was 3 (E1V1M1). Plain computed tomography scans showed a large hematoma with obstructive hydrocephalus in the left cerebellar hemisphere [Figure 1], and digital subtraction angiography showed the nidus of an AVM in the left cerebellar hemisphere with the left PPHA. The left PPHA was branching from the left internal carotid artery at the level of the C2 vertebral body. The left PPHA is connected to the basilar artery [Figure 2]. Upon admission, the intracranial pressure and vital signs were stabilized with emergency external ventricular drainage. The AVM in the left cerebellar hemisphere then was removed using an infratentorial-supracerebellar approach. The nidus, which was fed by the left supracerebellar artery, left posterior cerebral artery, anterior inferior cerebellar artery, and left posterior inferior artery, was drained via the precentral cerebellar vein and a tentorial bridging vein. The tentorial bridging vein was not easy to dissect because it was positioned in front of the nidus.

An angiogram of the left PPHA 2 weeks after the operation revealed residual nidus [Figure 3], which was treated using γ-knife surgery [Figure 4].

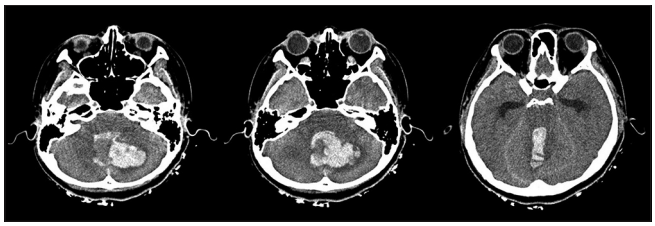

Figure 1: An initial computed tomography (CT) scan revealed a large hematoma and an acute obstructive hydrocephalus in the left cerebellar hemisphere

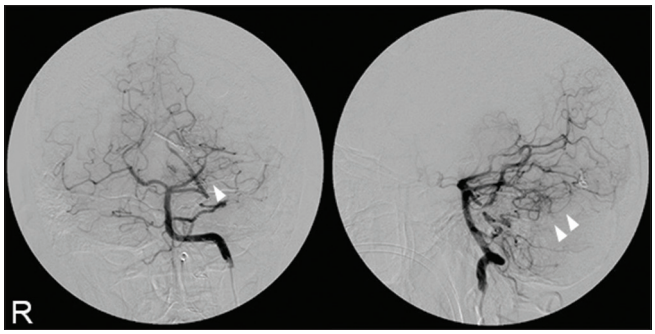

Figure 3: Digital subtraction angiography of the basilar artery (BA) via the PPHA (left, anteroposterior view; right, lateral view) after the surgery revealed the residual nidus (arrow heads)

DISCUSSION

Intracranial AVMs and CBAs are congenital vascular anomalies. AVMs develop during the late somatic stages of the 4th week of embryonic life and almost no later than the 8th week.[24] In contrast, CBAs disappear as the posterior communicating arteries develop by the 6th week of embryonic life.[7]

We hypothesized that the development of infratentorial AVMs disrupt the ability of CBAs to close.

To our knowledge, 28 AVMs associated with CBA have been reported in the English literature;[1-7,9-14,16,18,20-22,25-32] of those 28 cases, 5 (17.9%), including our case, were infratentorial AVMs [Table 1]. As infratentorial AVMs

Table 1: Summary of the 5 cases of infratentorial AVM associated with CBA

Authors	Age/sex	CBA	Site of AVM
Perret *et al.*[6]	Unknown	rt. PPTA	rt. side of pons
Nakai *et al.*[21]	58/M	lt. PPHA	lt. cerebellum
Ohtakara *et al.*[23]	21/F	lt. PPTA	lt. cerebellum
Yamanaka *et al.*[31]	46/M	lt. PPTA	lt. cerebellum
Present case	18/F	lt. PPHA	lt. cerebellum

CBA: Carotid-basilar anastomosis, AVM: Arteriovenous malformation; PPTA: Persistent primitive trigeminal artery; PPHA: Persistent primitive hypoglossal artery.

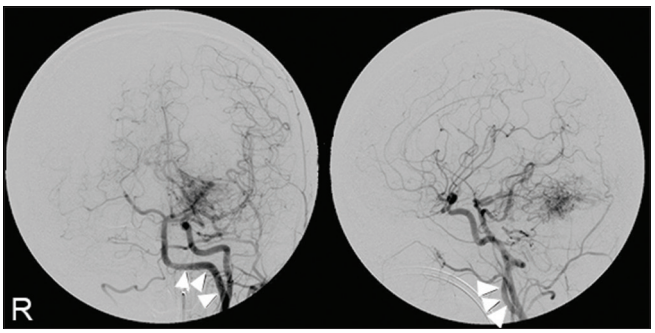

Figure 2: Digital subtraction angiography of the left common carotid artery (left, anteroposterior view; right, lateral view) showing the nidus of an arteriovenous malformation in the left cerebellar hemisphere and the left persistent primitive hypoglossal artery (PPHA; arrowheads)

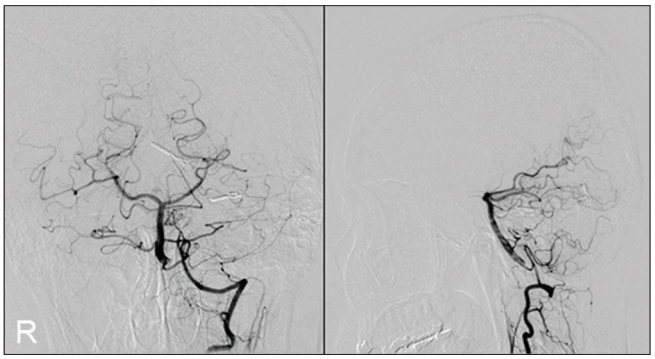

Figure 4: Digital subtraction angiography of the BA via the left vertebral artery (left, anteroposterior view; right, lateral view) 5 years after the γ-knife surgery. The residual nidus had disappeared

account for 6-8% of all AVMs,[8,15] the incidence of infratentorial AVM with CBA in these 28 cases of AVM associated with CBA is 2-3 times higher than that of common infratentorial AVM. In addition, all five infratentorial AVMs were located in the cerebellar hemisphere ipsilateral to the CBAs.

These findings strongly indicate a close relationship between infratentorial AVMs and persistent CBAs including PPHA. Embryologically, development of infratentorial AVM precedes the spontaneous closure of CBA. Once an AVM develops, a large amount of blood flows into the adjoining AVM via the ipsilateral vertebrobasilar system including the CBA, and this excess hemodynamic stress is a burden on the CBA. As a result, the spontaneous closure of the ipsilateral CBA may be disturbed. On the contrary, contralateral CBAs are never involved with the blood supply to the infratentorial AVM because bilateral CBAs are independent of each other until the longitudinal neural arteries merge to form the basilar artery at 31 days of embryonic life.[13,23]

CONCLUSION

Preexistence of an infratentorial AVM may induce formation of an ipsilateral persistent CBA; thus, coexistence of these two congenital anomalies is not incidental but inevitable.

REFERENCES

1. Agnoli AL. Vascular anomalies and subarachnoid haemorrhage associated with persisting embryonic vessels. Acta Neurochir (Wien) 1982;60:183-99.
2. Campbell RL, Dyken ML. Four cases of carotid-basilar anastomosis associated with central nervous system dysfunction. J Neurol Neurosurg Psychiatry 1961;24:250-3.
3. Fields WS. The significance of persistent trigeminal artery. Carotid-Basilar anastomosis. Radiology 1968;91:1095-101.
4. Gannon WE. Malformation of the brain. Persistent trigeminal artery and arteriovenous malformation. Arch Neurol 1962;6:496-8.
5. Garza-Mercado R, Cavazos E, Urrutia G. Persistent hypoglossal artery in combination with multifocal arteriovenous malformations of the brain: Case report. Neurosurgery 1990;26:871-6.
6. Harada K, Uozumi T, Kurisu K, Sumida M, Nakahara A, Migita K. Evaluation of cerebro-vascular diseases with persistent carotid-basilar anastomosis. No Shinkei Geka 1994;22:1153-8.
7. HD G, editor. Intracranial arteriovenous malformatons. 3rd ed. New York: McGraw-Hill; 1996.
8. Hernesniemi JA, Dashti R, Juvela S, Vaart K, Niemela M, Laakso A. Natural history of brain arteriovenous malformations: A long-term follow-up study of risk of hemorrhage in 238 patients. Neurosurgery 2008;63:823-9.
9. Jayaraman A, Garofalo M, Brinker RA, Chusid JG. Cerebral arteriovenous malformation and the primitive trigeminal artery. Arch Neurol 1977;34:96-8.
10. Karasawa JK, Furuse S, Kawai S, Uchida Y, Sakaki T. Studies on 2 cases of a persistent primitive hypoglossal artery. Case reports on a 9-year-old male patient with arteriovenous malformation and a 55-year-old male patient with brain stem syndrome. No To Shinkei 1973;24:91-8.
11. Karasawa J, Terano M, Nishikawa M, Kyoi K, Kikuchi H. Case of persistent bilateral carotid-basilar anastomoses (primitive otic artery and primitive trigeminal artery) with multiple vascular anomalies. No To Shinkei 1972;24:91-8.
12. Lamb J, Morris L. The carotid-basilar artery. A report and discussion of five cases. Clin Radiol 1961;12:179-86.
13. Luh GY, Dean BL, Tomsick TA, Wallace RC. The persistent fetal carotid-vertebrobasilar anastomoses. AJR Am J Roentgenol 1999;172:1427-32.
14. GÜNER MS. Arteriovenous malformation and persistent trigeminal artery association; Case report Turk Neurosurg 1996;6:33-6.
15. Moody RA, Poppen JL. Arteriovenous malformations. J Neurosurg 1970;32:503-11.
16. Mori HA, Maruyama K, Shin M, Tago M, Matsumoto T, Yoshikawa T, *et al*. Parietal arteriovenous malformation associated with ipsilateral persistent primitive hypoglossal artery. Riv Neuroradiol 2002;15:769-72.
17. Mullan S, Mojtahedi S, Johnson DL, Macdonald RL. Embryological basis of some aspects of cerebral vascular fistulas and malformations. J Neurosurg 1996;85:1-8.
18. Nakai Y, Yasuda S, Hyodo A, Yanaka K, Nose T. Infratentorial arteriovenous malformation associated with persistent primitive trigeminal artery--case report. Neurol Med Chir (Tokyo) 2000;40:572-4.
19. Nakajima K, Ito Z, Hen R, Uemura K, Matsuoka S. Congenital anomalies of cerebral artery and intracranial aneurysm. No To Shinkei 1976;28:197-201.
20. Nishida C, Ashikaga R, Araki Y, Nakamatsu K, Ono Y, Fujii K, *et al*. Persistent hypoglossal artery associated with arteriovenous malformation: A case report. Eur J Radiol 2000;33:59-62.
21. Ohtakara K, Kuga Y, Murao K, Kojima T, Taki W, Waga S. Posterior fossa arteriovenous malformation associated with persistent primitive trigeminal artery--case report. Neurol Med Chir (Tokyo) 2000;40:169-72.
22. Oran I, Parildar M, Memis A, Yunten N. Catheter and MR angiography of persistent trigeminal artery associated with occipital arteriovenous malformation. Comput Med Imaging Graph 2000;24:33-5.
23. Pasco A, Papon X, Bracard S, Tanguy JY, Ter Minassian A, Mercier P. Persistent carotid-vertebrobasilar anastomoses: How and why differentiating them? J Neuroradiol 2004;31:391-6.
24. Perret G, Nishioka H. Report on the cooperative study of intracranial aneurysms and subarachnoid hemorrhage. Section VI. Arteriovenous malformations. An analysis of 545 cases of cranio-cerebral arteriovenous malformations and fistulae reported to the cooperative study. J Neurosurg 1966;25:467-90.
25. Purkayastha S, Jayadevan ER, Gupta AK, Bodhey NK. Cerebral Arteriovenous Malformation Embolized through Persistent Primitive Hypoglossal Artery.A Case Report. Interv Neuroradiol 2005;11:392.
26. Sahoo YM, Mallikarjunaiah GS, David BS, Parida S. Persistent hypoglossal artery. Indian J Radiol 1988;42:144-6.
27. Shibata Y, Hyodo A, Saito A, Yoshii Y, Nose T. Large arteriovenous malformation associated with persistent primitive hypoglossal artery - case report. Neurol Med Chir (Tokyo) 1991;31:804-8.
28. Tomsick TA, Lukin RR, Chambers AA. Persistent trigeminal artery: Unusual associated abnormalities. Neuroradiology 1979;17:253-7.
29. Uchino A, Matsunaga M, Ohno M. Arteriovenous malformation of the corpus callosum associated with persistent primitive trigeminal artery-case report. Neurol Med Chir (Tokyo) 1989;29:429-32.
30. Wakai S, Ueda Y, Nagai M. Cerebrovascular anomalies associated with primitive trigeminal artery-report of three cases. Neurol Med Chir (Tokyo) 1987;27:633-6.
31. Yamanaka K, Noguchi K, Hayasaki K, Matsuoka Y. Persistent primitive hypoglossal artery associated with arteriovenous malformation-case report. Neurol Med Chir (Tokyo) 1990;30:949-55.
32. Yamauchi Y, Aoyama T, Sugitani Y, Koama M, Nukada T. Persistent hypoglossal artery associated with arteriovenous malformation-a study on cerebral circulation. Rinsho Shinkeigaku 1974;14:539-47.