Case Report

Infratentorial arteriovenous malformation associated with persistent primitive hypoglossal artery

Hiroshi Kageyama, Terushige Toyooka, Hideo Osada, Nobusuke Tsuzuki

Departments of Neurosurgery, Kuki General Hospital, 418-1 Kamihayami Kuki, National Defense Medical College, Namiki 3-2, Tokorozawa, Saitama, Japan

E-mail: *Hiroshi Kageyama - kageyamahiroshi29@gmail.com; Terushige Toyooka - teru_toy1809@friend.ocn.ne.jp; Hideo Osada - osah@ndmc.ac.jp; Nobusuke Tsuzuki - ntsuzuki0813@gmail.com

*Corresponding author

Received: 21 January 15  Accepted: 02 Mar 15  Published: 07 May 15

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

INTRODUCTION

Arteriovenous malformations (AVMs) are congenital vascular anomalies that arise as a result of abnormal blood vessel development during the early embryonic period. Of all AVMs, infratentorial AVMs account for 6-8%. 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). The angiographic incidence of PPH and PPTA are 0.05% and 0.20%, respectively.

To the best of our knowledge, the incidence of infratentorial AVM with CBA is 2-3 times higher than that of common infratentorial AVM. In addition, all reported infratentorial AVMs were
located in the cerebellar hemisphere ipsilateral to the CBA.\textsuperscript{[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 $\gamma$-knife surgery [Figure 4].

**DISCUSSION**

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

| Authors         | Age/sex | CBA     | Site of AVM  |
|-----------------|---------|---------|--------------|
| Perret et al.\textsuperscript{[6]} | Unknown | rt. PPTA | rt. side of pons |
| Nakai et al.\textsuperscript{[21]} | 58/M    | lt. PPHA | lt. cerebellum |
| Ohtakara et al.\textsuperscript{[23]} | 21/F    | lt. PPTA | lt. cerebellum |
| Yamanaka et al.\textsuperscript{[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.
account for 6-8% of all AVMs,\cite{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.\cite{11,23}

**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.

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