Large Arteriovenous Malformation Associated with Persistent Primitive Hypoglossal Artery

—Case Report—

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Abstract

A rare case of large arteriovenous malformation (AVM) with persistent primitive hypoglossal artery in a 43-year-old male was treated by staged embolization, followed by total removal. AVM associated with carotid-basilar anastomosis is quite rare, but the incidence of AVM in patients with carotid-basilar anastomosis is high. AVM associated with persistent carotid-basilar anastomosis has no distinguishing features compared with ordinary AVM, but participation of the posterior circulation as a feeder is characteristic.

Key words: arteriovenous malformation, carotid-basilar anastomosis, persistent primitive hypoglossal artery, embolization

Introduction

Primitive hypoglossal artery (PHA) is a rarely occurring persistent embryonal artery possibly associated with vascular anomalies such as aneurysm and arteriovenous malformation (AVM).¹⁾ Here we describe a case of a large AVM associated with persistent PHA, treated by staged embolization and total removal. We discuss reported cases of AVM with persistent carotid-basilar anastomosis, and the possible causes of the association.

Case Report

A 43-year-old male suffered sudden onset of a severe headache with nausea in the early morning of April 5, 1989. Computed tomographic (CT) scans on admission to another hospital suggested a large AVM in the left occipital region, but no obvious intracranial hemorrhage. However, the clinical symptom strongly suggested slight subarachnoid hemorrhage. He was transferred to our hospital for examination of the intracranial lesion on May 18, 1989.

On admission, neurological and physical examinations were normal except for cranial vascular bruit. CT scanning and magnetic resonance (MR) imaging demonstrated a large cerebral AVM, about 6 cm in diameter, located mainly in the left occipital lobe and extending into the temporoparietal region (Figs. 1 and 2). Cerebral angiography revealed a large AVM fed by the left middle cerebral, posterior cerebral, occipital, and meningeal arteries, and

Fig. 1 Pre- (left) and postcontrast (right) CT scans demonstrating a large vascular mass, about 6 cm in diameter, in the left occipital lobe.

Fig. 2 T_1 - (left: TR/TE 600/20 msec) and T_2 -weighted (right: TR/TE 2500/80 msec) axial MR images showing the left occipital lesion as flow void.

Fig. 3 *upper*: Left carotid angiograms, anteroposterior (left) and lateral (right) views, showing the left occipital AVM fed by the left middle cerebral, posterior cerebral, occipital, and meningeal arteries. *lower*: Left internal carotid angiograms, lateral views, revealing the primitive hypoglossal artery (arrow) arising from the internal carotid artery at the C2 level.

drained mainly by the superficial cortical veins and a deep vein (Fig. 3 *upper*). Left internal carotid angiography demonstrated left PHA originating from the left internal carotid artery at the C2 level and ending at the distal left vertebral artery (Fig. 3 *lower*). The AVM was located distally to the left PHA.

Fig. 4 Left carotid angiograms, anteroposterior (left) and lateral (right) views. *upper*: Angiograms after embolization showing about 90% of the initial AVM is embolized. *lower*: Angiograms after operation disclosing total removal of the AVM.

The AVM was embolized preoperatively because of the size and high flow. Ethylenevinyl alcohol and polyvinyl alcohol particles were infused transfemorally through a Tracker-18 infusion catheter (Target Therapeutics, San Jose, Cal., U.S.A.) and a leak balloon catheter (Dow Corning, Kanagawa). The procedure was repeated four times between June 7 and July 10 under local anesthesia with neurological examination and electroencephalographic monitoring. Superselective cannulation to the left posterior cerebral artery was performed *via* the left PHA. Right homonymous hemianopsia and temporally mild dyscalculia occurred after embolization. Cerebral angiography showed that about 90% of the initial AVM was embolized (Fig. 4 *upper*). The AVM was totally removed with relatively little bleeding on July 12, 1989.

Mild dyscalculia and dysgraphia appeared postoperatively. The right homonymous hemianopsia remained unchanged. Postoperative cerebral angiography confirmed the total removal of the AVM (Fig. 4 *lower*). His neurological deficits improved gradually. He was discharged on August 3, 1989, and resumed work as a farmer.

Table 1 Summary of reported cases of AVM with carotid-basilar anastomosis

Author (Year)	Age, Sex	Embryonal artery	Site of AVM	Main feeder	Participation of posterior circulation	Symptom	Therapy
Krakenbühl and Yasargil (1957) ¹³⁾	48, M	rt PTA	rt cerebellum	VA	+	neuralgia, vertigo	
Schiefer and Walter (1959) ²¹⁾	51, M	PTA	lt P			bleeding	extirpation
Weickmann (1959) ²⁷⁾	18, F	rt PTA	lt cerebellum	VA	+	bleeding	extirpation
Campbell and Dyken (1961) ²⁾	38, F	rt PTA	lt TP	MCA	—	epilepsy	
Lamb and Morris (1961) ¹⁴⁾	23, F	lt PTA	lt O	MCA, PCA	+	bleeding	extirpation
Gannon (1962) ⁸⁾	46, F	rt PTA	rt PO			bleeding	extirpation
Frugoni <i>et al.</i> (1963) ⁷⁾	18, M	rt PTA	rt PO	MCA, PCA	+	bleeding	
Eadie <i>et al.</i> (1964) ³⁾	?	PTA				bleeding	
	?	PTA				epilepsy	
Endo <i>et al.</i> (1965) ⁴⁾	?	rt PTA	infratentorial	VA	+		rt carotid ligation
Perret and Nishioka (1966) ¹⁹⁾	?	rt PTA	rt pons	PTA	+	bleeding	rt carotid trapping
Fields (1968) ⁶⁾	25, F	lt PTA	rt PO	MCA	—	bleeding	
Moody and Poppen (1970) ¹⁷⁾	?	PTA		PTA	+		
Karasawa <i>et al.</i> (1972) ¹²⁾	28, F	lt POA, rt PTA	great vein of Galen	PCA	+	bleeding	
Karasawa <i>et al.</i> (1973) ¹¹⁾	9, M	rt PHA	rt P	ACA, MCA	—	bleeding	preoperative death
Yamauchi <i>et al.</i> (1974) ²⁹⁾	51, M	lt PHA	rt P	ACA	—	bleeding	
Ferrante and Lorenzo (1976) ⁵⁾	28, M	lt PTA	rt PO			epilepsy	
Jayaraman <i>et al.</i> (1977) ¹⁰⁾	27, F	lt PTA	lt T	MCA, PCA	+	bleeding	conservative
Tomsick <i>et al.</i> (1979) ²⁴⁾	?	lt PTA	rt T	MCA	—	bleeding	
	17, M	rt PTA, rt POA	rt T	MCA, PCA	+	epilepsy	
Agnoli (1982) ¹⁾	43, M	rt PTA	rt O	PCA	+	epilepsy	
Wakai <i>et al.</i> (1987) ²⁶⁾	20, F	rt PTA	rt PO			bleeding	extirpation
Sahoo <i>et al.</i> (1988) ²⁰⁾	2, M	rt PHA	rt O	ECA, PHA	+	epilepsy	feeder ligation
Uchino <i>et al.</i> (1989) ²⁵⁾	16, F	lt PTA	corpus callosum	ACA, PCA	+	bleeding	irradiation
Mercado <i>et al.</i> (1990) ¹⁶⁾	30, F	rt PHA	lt P	MCA, AChA	—	bleeding	extirpation
Yamanaka <i>et al.</i> (1990) ²⁸⁾	46, M	lt PHA	lt cerebellum	PICA, SCA	+	bleeding	extirpation
Present case	43, M	lt PHA	lt O	MCA, PCA	+	bleeding (?)	embolization and extirpation

ACA: anterior cerebral artery, AChA: anterior choroidal artery, ECA: external carotid artery, MCA: middle cerebral artery, O: occipital lobe, P: parietal lobe, PCA: posterior cerebral artery, PHA: primitive hypoglossal artery, PICA: posterior inferior cerebellar artery, PO: parieto-occipital lobe, POA: primitive otic artery, SCA: superior cerebellar artery, T: temporal lobe, TP: temporoparietal lobe, VA: vertebral artery.

Discussion

Krakenbühl and Yasargil¹³⁾ reported the first case of AVM associated with persistent primitive trigeminal artery in 1957. Since then only 27 cases of AVM with carotid-basilar anastomosis have been reported (Table 1).^{1-8,10-14,16,17,19-21,24-29)} There was no significant sex predominance. The age at diagnosis ranged from 2 to 51 years old, but mainly between 20 and 40 years old. AVMs with carotid-basilar anastomosis vary in location, and are distributed similarly to general AVMs. The AVM was ipsilateral to the carotid-basilar anastomosis in slightly more cases than contralateral. Twenty-one of 27 cases included a feeding artery, and 15 of these cases had posterior circulation participation. The most frequent clinical

manifestation was intracranial hemorrhage.

Carotid-basilar anastomosis is frequently associated with aneurysm.^{1,14)} However, the significance of AVM associated with carotid-basilar anastomosis is controversial. Some authors consider it to be a chance one only,^{3,6)} but Wakai *et al.*²⁶⁾ showed a statistically higher incidence of vascular anomalies, *i.e.*, AVM and aneurysm, in patients with primitive trigeminal artery than in the general population.¹⁵⁾

An AVM probably originates at the second stage of Streeter when the primordial blood vessel plexus resolves into veins, arteries, and capillaries.^{9,18,23)} The vertebrobasilar system and posterior communicating artery finish development and the primitive carotid-basilar anastomosis regresses and disappears at the fifth stage.^{7,23)} Any effect of an AVM on the per-

sistence of the carotid-basilar anastomosis is unknown. However, an AVM may disturb the regression of the carotid-basilar anastomosis because the AVM requires a high blood flow. In our case, the coexistence of the AVM and the carotid-basilar anastomosis may have been associated, because the AVM was distal to the PHA. Seventy-one percent of such AVMs associated with carotid-basilar anastomosis were fed by the posterior circulation, a greater incidence than among general AVMs, which supports this hypothesis.

Our case was successfully treated with staged embolization followed by total removal. A large AVM has an especially high risk of normal perfusion pressure breakthrough (NPPB) following total removal or sudden embolization of feeders. Staged embolization followed by removal gradually changed the cerebral hemodynamics avoiding NPPB. Extensive AVM embolization assisted surgical removal by reducing intraoperative bleeding which may limit total excision.²²⁾

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