Arteriovenous Malformation with an Occlusive Feeding Artery Coexisting with Unilateral Moyamoya Disease

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Background　Arteriovenous malformations (AVMs) with vascular abnormalities, including aneurysms, have been reported frequently. However, the coexistence of AVM and unilateral moyamoya disease is rare. We report herein an AVM patient who presented with acute ischemic stroke with unilateral moyamoya disease and occlusion of the feeding artery.

Case Report　A-41-year old man was admitted with sudden dysarthria and facial palsy. Brain computed tomography and magnetic resonance imaging revealed an acute infarction adjacent to a large AVM in the right frontal lobe. Cerebral angiography revealed occlusions of the proximal right middle cerebral and proximal anterior cerebral arteries, which were the main feeders of the AVM. Innumerable telangiectatic moyamoya-type vessels between branches of the anterior cerebral artery and dilated lenticulostriate arteries on the occluded middle cerebral artery were detected. However, a nidus of the AVM was still opacified through the distal right callosomarginal artery, which was supplied by the remaining anterior cerebral artery and leptomeningeal collaterals from the posterior cerebral artery.

Conclusions　While AVM accompanied by unilateral moyamoya disease is rare, our case suggests an association between these two dissimilar vascular diseases.　**J Clin Neurol 2010;6:216-220**

Key Words　arteriovenous malformations, moyamoya disease, ischemic stroke.

Introduction

Arteriovenous malformation (AVM) is a vascular conglomerate known as a nidus which has a main arterial feeder that is connected directly to the draining veins. The incidence of AVM is below 1 : 100,000 and accounts for about 1-2% of all strokes, and 3% of those in young patients.[1] Although the association between AVMs and aneurysms is well known, an AVM in combination with moyamoya disease or progressive arterial occlusion has been reported only rarely.[2]

AVMs are generally thought to be congenital vascular anomalies that arise as a result of the abnormal development of blood vessels during the early embryonic period.[3] Conversely, moyamoya disease is a progressive vascular disease that is defined as bilateral distal occlusion or stenosis of the internal carotid artery (ICA) accompanied by a telangiectatic connection in the basal ganglia, the so-called 'moyamoya vessels'.[4] In particular, unilateral involvement is considered to be probable moyamoya disease. The etiology of vascular pathogenesis such as uni-lateral stenosis and occlusion is unknown, although variable secondary causes of arterial pathogenesis have been described, including atherosclerosis, brain tumor, and radiation.

We report herein a case of AVM with a feeding arterial occlusion and unilateral moyamoya disease coexisting in an acute stroke patient.

Case Report

A 41-year-old man was admitted with sudden dysarthria and left facial palsy. He had no history of hypertension, diabetes, hyperlipidemia, or smoking. Brain computed tomography (CT) demonstrated slightly hyperdense cortical lesions in the right frontal lobe with calcified foci and adjacent low-density ischemic lesions. CT angiography revealed occlusion of the right middle cerebral artery (MCA). T2-weighted magnetic resonance image (MRI) revealed the tightly packed vascular signals of flow voids without thrombus in the frontal lobe, and an adjacent ischemic lesion was confirmed by diffusion-weighted MRI.

No evidence of previous hemorrhagic lesions was detected on either the CT or MRI (Fig. 1). Digital subtraction angiography revealed occlusion of the right proximal callosomarginal artery, a branch of anterior cerebral artery (ACA), and many small moyamoya-type telangiectatic connections between the mid callosomarginal and the pericallosal arteries. Occlusion of the right proximal MCA and dilated lenticulostriate arteries were also seen during the arterial phase. The right superior branches of the MCA and an AVM nidus were drained into the posterior cerebral artery-leptomeningeal collateral connection during the capillary-venous phase. However, no aneurysm was detected in the angiographic study (Fig. 2). Moreover, we failed to find any cardioembolic or atherothrombotic source in the electrocardiogram, carotid Doppler, or transthoracic echocardiography. Laboratory testing revealed normal levels and activities of protein C, S antigen, antiphospholipid antibody, anticardiolipin antibody, rheumatoid factor, fluorescent antinuclear antibody, lupus erythematosus cell, and homocysteine. Surgical interventions were not performed on the patient due to the mild symptoms and the presence of abundant collateral vessels. The patient was discharged without major disability on the 8th day of hospital treatment.

Discussion

Our patient exhibited coexistence of two rare arterial lesions: 1) an AVM with proximal callosomarginal artery occlusion, a feeder of the AVM, and 2) unilateral MCA occlusion with basal moyamoya vessels. The patient presented no evidence of atherosclerosis, inflammation, radiation, or trauma. Acute cerebral lesions in the adjacent AVM were the result of occlusion of the MCA. Abundant telangiectatic connections between the callosomargianal and pericallosal arteries were revealed with angiography.

AVMs may be accompanied by dysplastic vascular changes including occlusion, stenosis, aneurysms, and ectasis. The rate of coexistence of aneurysm and AVMs in the same patient has been reported to be up to 58%. However, progressive feeding-artery occlusion of the AVM has been detected in less than 3% of cases.[2,5] The finding of an AVM accompanied by moyamo-
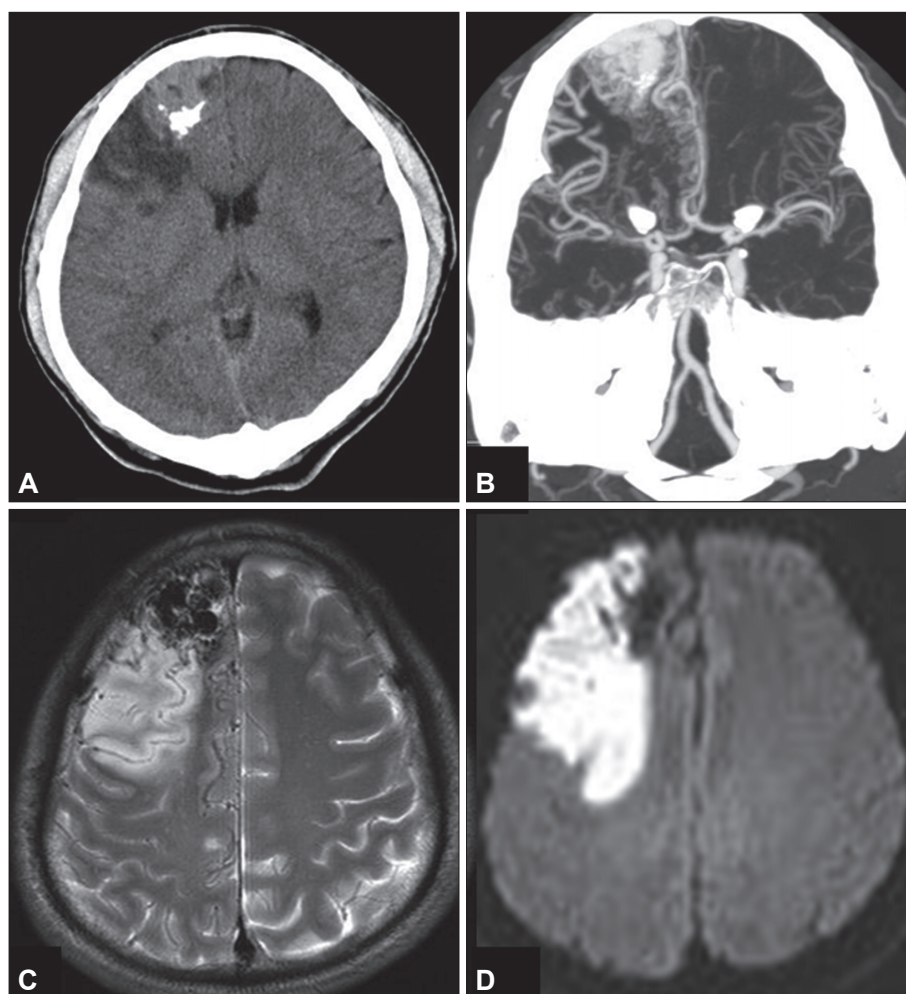
Fig. 1. Results of brain computed tomography (CT) and magnetic resonance imaging (MRI). A: Pre-contrast brain CT scan showing hypodense lesions and a slightly hyperdense area with an irregular calcified lesion in the right frontal cortex. B: Coronal CT angiography showing enlarged draining veins and proximal right middle cerebral arterial occlusion. C: T2-weighted MRI, flow-void signals in the right frontal area. D: Diffusion-weighted MRI showing high signal intensity lesion in the right anterior and middle cerebral arterial territories.
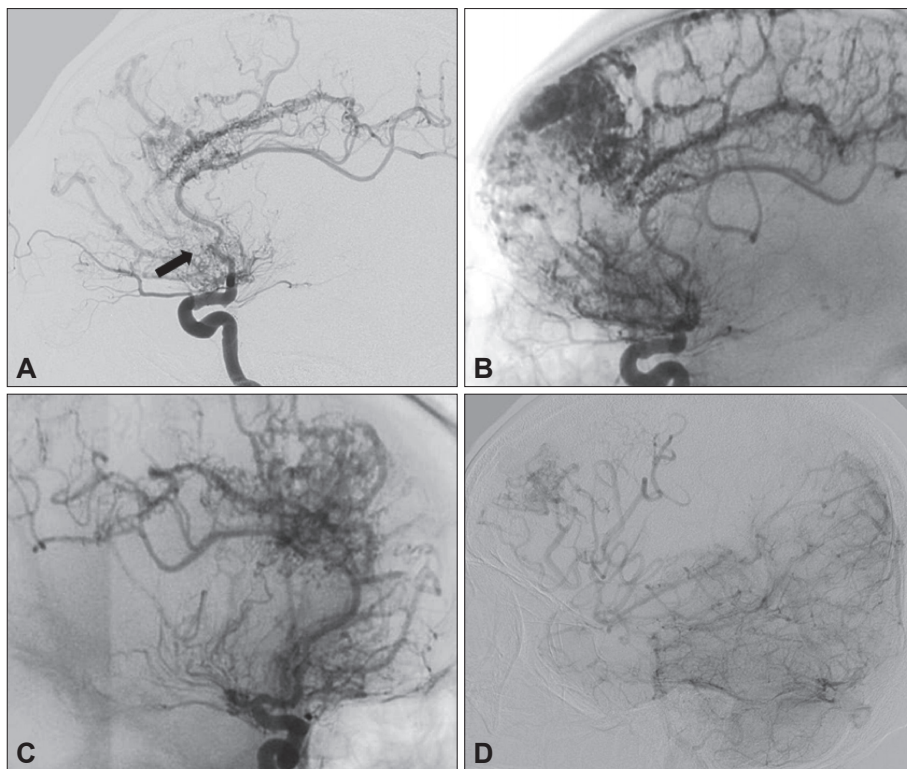
Fig. 2. Digital subtraction angiography. A: Lateral early arterial phase of the internal carotid artery shows the callosomarginal artery, which is occluded proximally (arrow), reappeareing in the distal part through abundant telangiectatic connections from the pericallosal artery. B: A nidus with dilated cortical draining veins was detected in the late arterial phase. C: Occlusion of the proximal middle cerebral artery with prominent moyamoya vessels evident in the right anterior oblique view. D: Distal branches of the middle cerebral artery are opacified from anterotemporal and prieto-occipital branches of the right posterior cerebral artery in the capillary phase of lateral vertebral angiography.

ya disease is even more rare. Only 21 cases have been reported in the English literature, as revealed by a scanning of the PubMed search and related articles (Table 1). The mean age of these patients was 32.1 years (range 3-54 years) with no gender predilection. Clinical presentation resulted in ischemic symptoms or infarction in 13 patients, intracranial hemorrhage in 7, and headache in 1. The mean age of patients with hemorrhagic symptoms was greater than that of those with ischemic symptoms (38.3 and 28.1 years, respectively). Most ischemic presentations seemed to be associated with moyamoya disease, because ischemic stroke in AVM patients is rare feature.

It is unknown which vascular anomaly developed first: the AVM or moyamoya disease. Most reported cases show that AVMs located in the ipsilateral side of moyamoya vessels are associated with a moyamoya-type collateral.[6-16] One hypothesis is that vascular occlusion drives from acquired dysplasia as a consequence of high blood flow from the AVM. The clinical evidence of high-flow-induced vascular change can be found in two case series of 19 patients with AVMs with progressive stenosis or occlusion of the feeding artery.[5,17] Enam and Malik reported that six patients had a AVM with unilateral ICA occlusion with proximal ACA or MCA occlusions, and one had a cerebellar AVM with proximal occlusion of the vertebral artery.[5] Mawad et al.[17] reported nine AVM patients with moyamoya-type collaterals and three patients with isolated MCA or ACA occlusion without collaterals. In all cases, occlusive arterial lesions were observed in the proximal artery that supplied the

AVM and were located unilaterally on the ipsilateral side of AVM. AVMs with feeding-arterial occlusion were accompanied with abundant angiectatic capillaries or collaterals resembling those found in moyamoya disease. The authors assumed that increased blood flow through an AVM causes turbulent flow at the carotid artery bifurcation, leading to focal intimal hyperplasia, with subsequent ICA stenosis and development of the moyamoya-type collaterals.

Others have suggested that the AVM most likely occurs secondarily to the moyamoya disease. There are three case reports of an initial evaluation that failed to find AVM in a moyamoya patient, but where subsequent study discovered an AVM to be present.[18-20] Those authors assumed that the ischemic environment caused by moyamoya disease could influence the development of an AVM, since proximal large-arterial stenosis could be attributed to vasodilation in distal vessels, resulting in arterio-vascular shunts. Increased levels of basic fibroblast growth factor, which is a known angiogenic factor found in the endothelium or smooth muscle cells, have been detected in the cerebrospinal fluid of moyamoya disease patients.[21] These factors may lead to opening and distension of anastomotic channels from the moyamoya vessels to the cortical veins concurrently with hyperangiogenic environments, in combination with the local angiogenic stimulation of previously injured neural tissue. However, such AVMs were small (less than 3 cm), and were filled with contrast dye during the late rather than the early arterial phase, which differs from the case in typical AVMs.

Another hypothesis is that the coexistence of AVMs and moyamoya disease is coincidental finding. In three cases, moyamoya vascular changes were found in locations unrelated to the AVM.[22-24] However, this differs from the cases mentioned above wherein the AVMs were supplied by normal feeding arteries, not by moyamoya collateral vessels.

Table 1. Summary of cases reported with an arteriovenous malformation (AVM) accompanied by moyamoya disease

References	Age/gender	Stenosis	AVM location	Clinical presentation	Treatment	Prognosis
Kayama et al, 1986	33/M	Bilateral ICA	Lt. temporo-occipital	Transient weakness, infarction	Carotid sympathectomy, ganglionectomy, EDAS	Symptom free
Lichtor & Mulan, 1987	34/M	Rt. ACA, MCA, Lt. ACA	Corpus callosum	Infarction	Conservative	Distal limb weakness
	50/F	Rt. ACA, MCA, Lt. ACA	Rt. parietal	Infarction	Rt. EDAS	Minimal Lt. arm weakness
	43/F	Rt. MCA, Lt. MCA, ACA	Rt. front-central	ICH	Revascularization	Lt. hemiplegia, aphasia
Okada et al, 1990	38/F	Bilateral ICA	Lt. frontal	Infarction	AVM resection, encephalo-aponeurotic synangiosis	Rt. hemiparesis, aphasia
Montanera et al, 1990	54/F	Bilateral ICA	Bifrontal	Transient numbness, dysphasia	Conservative	NA
	44/F	Rt. MCA	Rt. parietal	Transient weakness	Conservative	NA
Akiyama et al, 1994	42/F	Bilateral ICA	Lt. frontal	Rt. BG ICH, Seizure	Bilateral EMS	Residual hemiparesis
Fuse et al, 1996	5/F	Bilateral ICA	Lt. frontal	Headache	EDAS	Repeated headache
Shimit et al, 1996	3/M	Bilateral ICA, PCA	Lt. parietal	Infarction	Conservative	NA
Halatsch et al, 1997	30/F	Bilateral ACA, MCA	Lt. parietal	Ischemia	STA-MCA anastomosis, AVM resection	Recurrent ischemic stroke
Voros et al, 1997	38/F	Rt. ACA, MCA	Rt. frontal	Ischemia	STA-MCA anastomosis, AVM resection	NA
Nakashima et al, 1998	37/M	Bilateral ICA	Rt. occipital	Ischemia	EDAS	Hemiparesis
	44/F	Bilateral ICA	Lt. frontal	Left BG ICH	Conservative	Hemiparesis, aphasia
Seol et al, 2002	23/M	Bilateral ICA	Lt. frontal	Seizure, transient weakness	EDAS, gamma-knife surgery	Symptom free
Fasseett et al, 2004	44/M	Lt. MCA	Lt. parietal	SAH, ischemia symptom	AVM resection	Discharge with hand weakness
O'Shaughnessy et al, 2005	6/F	Rt. ICA	Rt. temporal	Infarction	AVM resection	Symptom free
Nwawai et al, 2006	35/M	Lt. ICA	Lt. parietal	Seizure, ICH	Conservative	Discharge with hemiparesis
Somasundaram et al, 2007	48/M	Bilateral ICA	Lt. frontal parasagittal	SAH	STA-MCA anastomosis	Discharge without deficit
Chen et al, 2007	8/M	Rt. ICA, bilateral PCA	Rt. BG, thalamus	Transient weakness	Conservative	NA
Deng et al, 2008	16/F	Rt. MCA	Lt. cerebellum	ICH	AVM resection	Discharge without deficit

M: male, F: female, Rt: right, Lt: left, ICA: internal carotid artery, ACA: anterior cerebral artery, MCA: middle cerebral artery, PCA: posterior cerebral artery, VA: vertebral artery, BG: basal ganglia, ICH: intracranial hemorrhage, EDAS: encephaloduroarteriosynangiosis, STA: superficial temporal artery, EMS: encephalomyosynagiosis, NA: not available.

Unilateral moyamoya disease, which involves occlusion or stenosis of the ICA or proximal MCA with moyamoya vessels, is atypical and is especially rare in adult. Some patients with unilateral moyamoya disease progress to the bilateral form. However, the low level of basic fibroblast growth factor in the CSF and lack of familial occurrence in unilateral moyamoya disease supported a different pathomechanism that is different from typical bilateral moyamoya disease.[25] Clinically, increased levels of atherogenic factors or high regional blood flow are reported in unilateral moyamoya disease.[26]

In our case, occlusion of the AVM feeder was detected in the right proximal ACA. During the injection of contrast dye into the vertebral artery, delayed contrast filling of right MCA from the leptomeningeal arteries supports the suggestion that the MCA was also a main feeder of the AVM. Well-developed basal and leptomeningeal collaterals were the result of slow progressive vascular pathology.[27] Therefore, high flow caused by the AVM may influence the development of unilateral moyamoya disease, and acute ischemic stroke is thought to be the result of progressive vascular occlusive disease. Although the simultaneous presence of two vascular diseases that have different characteristics is extremely rare, further study is necessary to improve our understanding of the association between AVMs and moyamoya disease.

Conflicts of Interest

The authors have no financial conflicts of interest.

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