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Mutsumasa Takahashi

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Magnification Angiography of Cerebral Aneurysms Associated with Moyamoya Disease

Mutsumasa Takahashi¹

The true cause of intracranial hemorrhage in moyamoya disease has not been clarified. Magnification cerebral angiograms were reviewed for seven patients. In four, small saccular aneurysms were seen arising from abnormally dilated collateral vessels or feeding arteries. This study suggests that aneurysms may be a source of hemorrhage in addition to the generally accepted rupture from abnormally dilated collateral vessels.

Most patients with moyamoya disease are seen initially with intracranial bleeding [1, 2]. Although dilated abnormal vascular networks in the base of the brain are reportedly responsible for the hemorrhages [3, 4], their true cause has not been elucidated [2, 3, 5]. Recently, the intracranial hemorrhages have been attributed to cerebral aneurysms, but the incidence of such aneurysms is relatively low [4, 6, 7]. Review of magnification angiograms of patients with moyamoya disease revealed that saccular aneurysms were more prevalent than previously reported. This report describes the angiographic features of moyamoya disease, with special attention to these associated aneurysms.

Materials and Methods

Magnification cerebral angiography was performed in six patients with moyamoya disease using 0.1×0.1 mm focal spots [8] with a magnification factor of 2.0–2.5. A series of nine or 10 films was obtained. In a seventh patient, nonmagnification cerebral angiography was performed. In each case, both internal and external carotid arteries and one of the vertebral arteries were selectively catheterized. Simultaneous biplane angiography was not performed.

Four of the seven patients had cerebral aneurysms, and each case met the criteria of moyamoya disease: stenosis or occlusion of the supraclinoid part of the internal carotid artery and the most proximal parts of the anterior and middle cerebral arteries with extensive abnormal vascular networks in the base of the brain. Diagnosis of aneurysms required good visualization on both lateral and anteroposterior views.

Seven other well documented cases of moyamoya disease with aneurysms were collected from the Japanese and English literature [4, 6, 7] and compared with those in my series. The size of the aneurysms was corrected for magnification.

Case Reports

Case 1

A 10-year-old girl developed gait and speech disturbances 6 months before admission. She experienced a left-sided clonic hemiconvulsion originating in the left arm 3 days before admission. Physical examination revealed a left hemiparesis with decreased sensation in the left upper extremity and left side of the body. Both lower extremities were unremarkable. There was slight muscular weakness on the left, especially in the left arm, but no evidence of muscular atrophy. A lumbar puncture was not performed.

Computed tomography (CT) of the brain demonstrated focal decreased x-ray absorption

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¹Department of Radiology, Akita University School of Medicine, 1-chome, Hondo, Akita 010, Japan.

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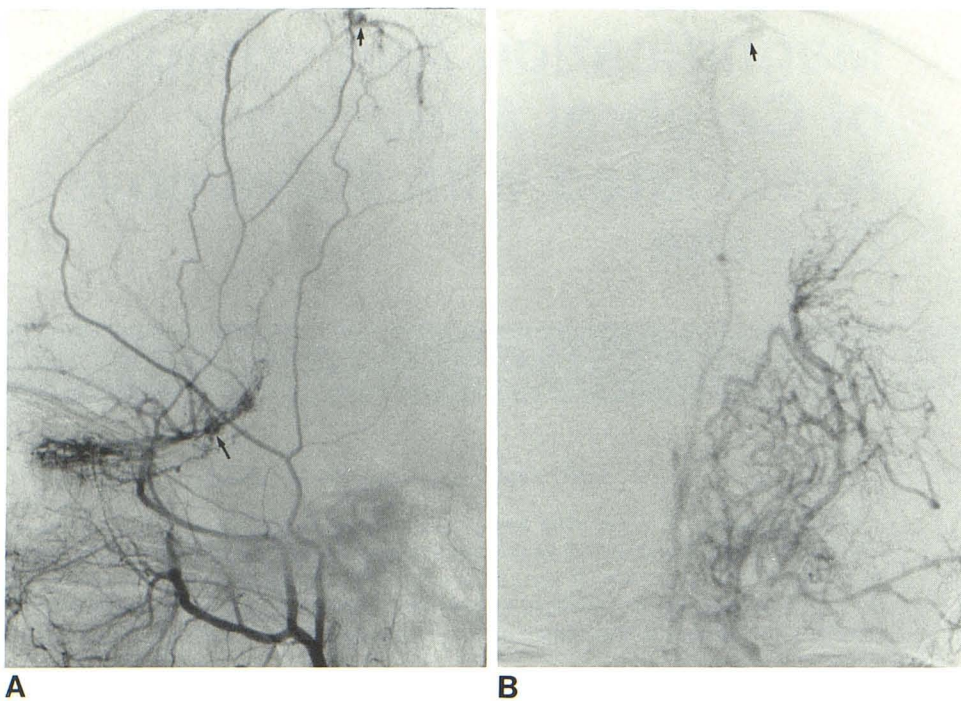


Fig. 1.—Case 1, 10-year-old girl with moyamoya disease. **A**, Left external carotid angiogram, lateral view. Aneurysm of 2 mm at junction of middle meningeal artery and branch of anterior cerebral artery near vertex (*short arrow*). In suprasellar area, 4 mm saccular aneurysm arises from abnormal artery between middle meningeal and internal carotid artery (*long arrow*). **B**, Left internal carotid angiogram, anteroposterior view. Extensive abnormal vascular networks at base of brain with partial visualization of medullary arteries. Small aneurysm on convexity medially at junction of middle meningeal artery and branch of anterior cerebral artery (*arrow*). Aneurysm at base of brain not demonstrated.



Fig. 2.—Case 2, 54-year-old woman with moyamoya disease. Vertebral arteriogram, Towne view. Saccular aneurysm of 8 mm arises at basilar apex, protruding superiorly.

in the right temporoparietal region, the right basal ganglia, and in the right frontal lobe due to old cerebral infarctions. Contrast enhancement revealed tortuous vessels in the basal ganglia; the circle of Willis and the insular vessels on the right side were not visualized.

Internal carotid angiography demonstrated occlusion of the su-

praclinoid parts of the internal carotid arteries bilaterally with extensive abnormal vascular networks and medullary collaterals. The horizontal part of the middle cerebral artery was completely occluded on both sides with reconstitution of the sylvian vessels via communications between the abnormal vascular networks and insular segments. On the left vertebral angiogram there was occlusion of the left parietooccipital and calcarine arteries with reconstitution of distal segments of these vessels via leptomeningeal collaterals. There were extensive collaterals connecting the posterior and anterior circulations via abnormal vascular networks.

A 2 mm saccular aneurysm arose from the anastomotic site between the left middle meningeal artery and a branch of the anterior cerebral artery in the frontoparietal area (fig. 1). Another 4 mm aneurysm was found in the suprasellar area arising from an abnormal vessel at the communication between the left middle meningeal and internal carotid artery.

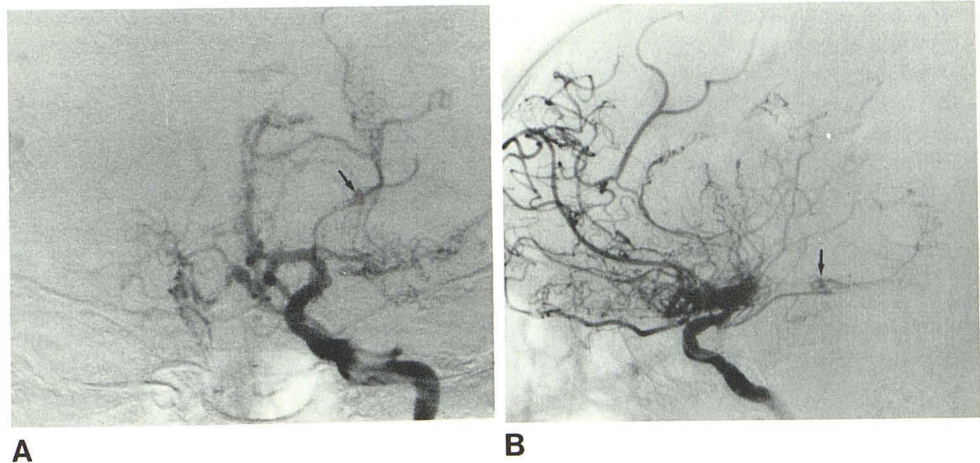
Case 2

A 54-year-old woman developed a left temporal headache with nausea and loss of consciousness. On admission 2 weeks later she was conscious and alert but had neck stiffness. A spinal tap revealed xanthochromia and Kernig sign was positive, but there was no increase in intracranial pressure. The deep tendon reflexes were normal and there was no pathologic reflexes.

Computed tomography demonstrated decreased x-ray absorption in the right frontal region and the left basal ganglia, with a large infarction in the right parietal lobe. There was minimal to moderate frontal atrophy and ventricular enlargement. Following infusion with contrast media there were tortuous vessels in the basal ganglia. The circle of Willis was partly visualized.

Cerebral angiography demonstrated typical occlusion and collateral vessels secondary to moyamoya disease. There was extensive anastomosis between the perforators and insular arteries on the left side. An 8 mm saccular aneurysm was noted at the basilar apex (fig. 2).

Fig. 3.—Case 3, 59-year-old man with moyamoya disease. **A**, Left internal carotid angiogram, anteroposterior view. Middle cerebral artery occluded at origin; anterior cerebral artery occluded; frontopolar artery functions as collateral vessel. Abnormal vascular networks formed at base of brain; collaterals arising from ophthalmic artery in ethmoid area. Saccular aneurysm of 2.5×3.5 mm arises from left anterior choroidal artery, probably within choroid fissure (arrow). **B**, Left internal carotid angiogram, lateral view. Abnormal vascular networks and collateral vessels in ethmoid region. Occluded middle cerebral and anterior cerebral arteries. Small aneurysm arises from anterior choroidal artery (arrow).



Case 3

A 59-year-old man had a headache and nausea with impaired consciousness $4\frac{1}{2}$ years before admission. A spinal tap revealed bloody cerebrospinal fluid and carotid angiography revealed moyamoya disease. He experienced vertigo, right hemiparesis, and right motor weakness again 3 weeks before admission. Physical examination revealed right motor weakness.

Computed tomography of the brain demonstrated areas of decreased x-ray absorption in the left temporal region and left basal ganglia. After infusion of contrast media, focal enhancement occurred in the low density lesions.

Carotid angiography demonstrated complete occlusion of the supraclinoid part of the right internal carotid artery and occlusion of the left middle cerebral artery at its origin from the internal carotid artery. There were extensive parenchymal and leptomeningeal collaterals, which reconstituted the middle cerebral arteries. Extensive parenchymal, transdural, and leptomeningeal collaterals were noted. The left parietooccipital and calcarine arteries were occluded on the left vertebral angiogram. Extensive leptomeningeal collaterals reconstituted these occluded parts. In addition, there were leptomeningeal collaterals toward the anterior circulation. A 3.5×2.5 mm saccular aneurysm arose from the left anterior choroidal artery, probably within the choroid fissure (fig. 3).

Case 4

An 8-year-old girl developed weakness of the left lower extremity 4 years before admission. This weakness recurred once or twice a month, but gradually resolved. At 2 years before admission she had severe headaches once or twice a month that resolved with medication. At 1 month before admission, she experienced severe and persistent headaches. Her eyesight was found to be impaired during a school eye examination. When evaluated elsewhere, her electroencephalogram was abnormal, but her spinal tap was unremarkable. Physical examination revealed no significant abnormality.

Bilateral carotid and left vertebral angiography revealed extensive abnormal vascular networks at the base of the brain, with occlusion of the supraclinoid parts of both internal carotid arteries and narrowing of the proximal anterior and middle cerebral arteries. The distal cortical branches of the anterior and middle cerebral arteries were reconstituted via abnormal vascular networks and transdural and leptomeningeal collaterals. There was also marked development of the medullary collaterals on both sides. The right posterior cerebral artery was unremarkable, but a 2×3 mm

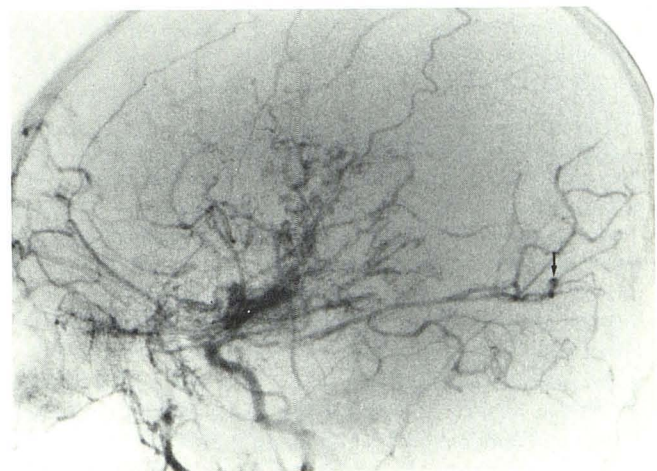


Fig. 4.—Case 4, 8-year-old girl with moyamoya disease. Left internal carotid angiogram, lateral view. Occlusion of supraclinoid part of internal carotid artery with extensive abnormal vascular networks and leptomeningeal collaterals. Saccular aneurysm of 2×3 mm arises from distal segment of left posterior cerebral artery.

saccular aneurysm arose from the distal segment of the left posterior cerebral artery (fig. 4).

Results

Eleven patients had aneurysms associated with moyamoya disease. Four were from my series and seven from the literature [4, 6, 7]. The group comprised six males and five females 8–59 years old.

All except the two pediatric patients (cases 1 and 4) had one or more episodes of intracranial bleeding. These two children had symptoms secondary to cerebral ischemia. One of my patients and one from the series of Kodama and Suzuki [7] had two aneurysms. The others all had single aneurysms.

Three aneurysms arose from the posterior choroidal ar-

tery; one arose from the anterior choroidal artery and one from the abnormal vascular networks. One aneurysm arose from the junction of the middle meningeal artery and a cortical branch. Four aneurysms arose from the basilar apex and one from the origin of the superior cerebellar artery.

Most aneurysms were saccular; fusiform aneurysms were rare. All the aneurysms were small (1.5–4.0 mm diam), except for the basilar artery aneurysms, the largest of which was 8 mm diam.

Discussion

The most common clinical manifestations of moyamoya disease are subarachnoid, intracerebral, or intraventricular hemorrhages [1, 2]. The incidence of hemorrhage exceeds 60% in adults and is 10% in children [2].

Intracranial bleeding in moyamoya disease has been attributed to the rupture of dilated collateral vessels or abnormal vascular networks [4]. Recently, cases with associated intracranial aneurysms were reported, and it has been suggested that the associated aneurysms may be another important cause of intracranial bleeding [4, 6, 7].

Associated intracranial aneurysms were recognized in four of seven of my cases and so far I have found seven other well documented cases in the literature [4, 6, 7]. These aneurysms usually arise from the anterior and posterior choroidal arteries, the basilar apex, and abnormally dilated collaterals, but other arterial branches and major arterial segments may also give rise to aneurysms. The aneurysms are usually small and saccular. The incidence of aneurysms associated with moyamoya disease is reported as 14% in adults [7]. I believe this figure is too low because intracranial bleeding occurs in more than 60% of adults with moyamoya disease. In two of my four patients with aneurysms a subarachnoid hemorrhage had occurred.

With routine nonmagnification angiography, small aneurysms may not be visualized due to the lower resolution and the overlapping vessels. Using selective angiography with magnification technique, cerebral aneurysms were discovered in four of seven cases. Furthermore, fusiform or saccular dilatations, which simulate aneurysms, were often visible in many arterial branches. Some cases of intracranial hemorrhage probably occur from microaneurysms that cannot be demonstrated angiographically or from abnormally dilated collaterals or feeding arteries, as has been reported previously [4].

It is difficult to postulate why the aneurysms associated with moyamoya disease arise frequently from the anterior and posterior choroidal arteries. Aneurysms are also com-

mon in the vertebrobasilar system. These arteries most often extensively supply and give rise to abnormal collateral vessels.

The pathogenesis of these aneurysms is unknown. Kodama and Suzuki [7] assumed that they were pseudoaneurysms arising at the sites of rupture and hemorrhage of small arterial branches. This conclusion was based on the fact that these aneurysms spontaneously regressed and completely resolved. Although it is difficult to explain this resolution, it may be because of arterial spasm secondary to subarachnoid hemorrhage. The arterial wall of these vessels also may undergo degenerative change and weakening, with a tendency to dilatation or rupture [4, 6]. This may produce saccular aneurysms. This mechanism is probably similar to that involved in the development of aneurysms in feeding arteries of arteriovenous malformations [9]. It is also possible that there are small congenital aneurysms in the collateral arteries or feeding vessels before moyamoya disease develops.

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