Mechanism of Moyamoya Vessels Secondary to Intracranial Atherosclerotic Disease: Angiographic Findings in Patients with Middle Cerebral Artery Occlusion

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Moyamoya vessels, which form a fragile collateral vascular network at the base of the brain, are associated with moyamoya disease and moyamoya syndrome. The mechanisms involved in their development remain unknown. This study evaluated the angiographic findings of the collateral vasculature in patients with moyamoya vessels secondary to atherosclerotic middle cerebral artery (MCA) occlusion. The study population comprised 26 patients with chronic atherosclerotic unilateral MCA occlusion who underwent digital subtraction angiography. We evaluated the presence of moyamoya vessels and intracranial arterial lesions and the degree of leptomeningeal anastomosis. We divided the patients into two groups, those with and those without moyamoya vessels, and compared clinical backgrounds and angiographic findings between the groups. Of the 26 patients, 17 had moyamoya vessels. The presence of moyamoya vessels was associated with ipsilateral or contralateral anterior cerebral artery stenosis to the occluded MCA ($P = .004$) and poor development of anterior leptomeningeal anastomosis ($P = .012$). It also was associated with vascular lesions involving more than one intracranial branch vessel in patients with moyamoya syndrome secondary to atherosclerosis. Our findings suggest that moyamoya vessels might be compensatory collateral vessels associated with poorly developed leptomeningeal collateral vessels. **Key Words:** Cerebrovascular disease—intracranial stenosis—cerebral artery—collateral circulation.

Moyamoya vessels, a network of dilated fragile collateral vessels at the base of the brain, are associated with moyamoya disease and moyamoya syndrome.[1,2] Moyamoya disease is characterized by progressive stenosis of the bilateral terminal portion of the internal carotid artery and its main branches, with no known cause. Moyamoya syndrome is a cerebrovascular occlusive disorder with similar angiographic findings to moyamoya disease and causes secondary to systemic diseases, such as atherosclerosis and inflammatory diseases. Several studies have suggested that moyamoya vessels are associated with hemodynamic impairment and compensatory collateral vasculature in moyamoya disease.[3,4] However, few studies have evaluated the moyamoya vessels in moyamoya syndrome, and the mechanism of development of these moyamoya vessels remains unknown. The aim of this study was to clarify the association between the collateral vasculature and the presence of moyamoya vessels. To do so, we evaluated the angiographic findings in patients with moyamoya vessels secondary to atherosclerotic middle cerebral artery (MCA) occlusion.

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Received August 31, 2010; accepted October 14, 2010.

Supported by a grant from the Research Committee on Moyamoya Disease established by the Ministry of Health, Labor and Welfare of Japan.

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1052-3057/$ - see front matter

doi:10.1016/j.jstrokecerebrovasdis.2010.10.001

Patients and Methods

Patients

A total of 30 patients with chronic MCA proximal occlusion on one side underwent cerebral digital

subtraction angiography (DSA) in our department between March 2000 and June 2010. Patients with cerebral artery occlusion of unknown origin (n = 3) and patients with cerebral angiitis (n = 1) were excluded from the present study, given our focus on moyamoya syndrome caused by atherosclerosis. The remaining 26 patients (15 men and 11 women, aged 38-74 years) were included in the study. None of the patients had undergone extracranial-to-intracranial arterial bypass surgery. Thirteen of the patients had been included in a previous study investigating hemodynamic and metabolic status in patients with MCA occlusion by positron-emission tomography (PET).[5] All patients demonstrated occlusion at the proximal M1 portion. No patient had stenosis in the terminal portion of the ipsilateral internal carotid artery. The cause of the MCA occlusion was clinically presumed to be atherosclerotic in all patients, based on the presence of atherosclerotic cerebrovascular changes in other areas and one or more atherosclerotic risk factors (Table 1). The study group included 20 symptomatic patients and 6 asymptomatic patients. The symptomatic patients had a history of transient ischemic attack (n = 9) or minor ischemic stroke (n = 11). None of the patients had a large infarct (>30 mm in diameter) evaluated by magnetic resonance imaging. Chronic occlusion was defined as an occlusion detected incidentally in an asymptomatic patient or an occlusion detected at least 1 month after the onset of symptoms in a symptomatic patient. All study protocols were approved by the Osaka University Hospital's Ethics Committee for Clinical Research.

Table 1. *Clinical background of the study subjects (n = 26)*

Age, years, mean ± SD	61.0 ± 10.8
Sex, M/F, n	15/11
Hypertension, n	21
Diabetes, n	7
Dyslipidemia, n	18
Current smoking, n	5
History of cerebrovascular disease, n	20
Ischemic stroke	11
Transient ischemic attack	9
Hemorrhagic stroke	0
Maximal infarct size, n	
No infarct	8
<15 mm	10
15-30 mm	8

Angiographic Assessment

All patients underwent DSA for clinical purposes, performed with an Integris V3000 unit (Philips Medical Systems, Best, The Netherlands). The results were first reviewed independently by two investigators (M.T. and M.S.) who were blinded to all clinical information about the patients. Following discussion, these two investigators reached a consensus on the angiographic findings for each patient.

The anatomic information evaluated included (1) the side and portion of the MCA occluded, (2) the presence of moyamoya-like vessels at the base of the brain on the occluded MCA side (Fig 1), (3) the presence of occlusion or stenosis in the anterior cerebral artery (ACA) or posterior cerebral artery (PCA), (4) the presence of the anterior communicating arteries, and (5) the segment of MCA in which retrograde contrast fillings from the ACA reached the occluded MCA side.[5] Sufficient leptomeningeal collateral vessels were defined as those that reached the M1 or M2 segment, and insufficient collateral vessels were defined as those that reached the M3 segment or the apparent absence of collateral vessels.

Given the considerable variability in the anatomy of the circle of Willis, a focal narrowing or occlusion of the vessel

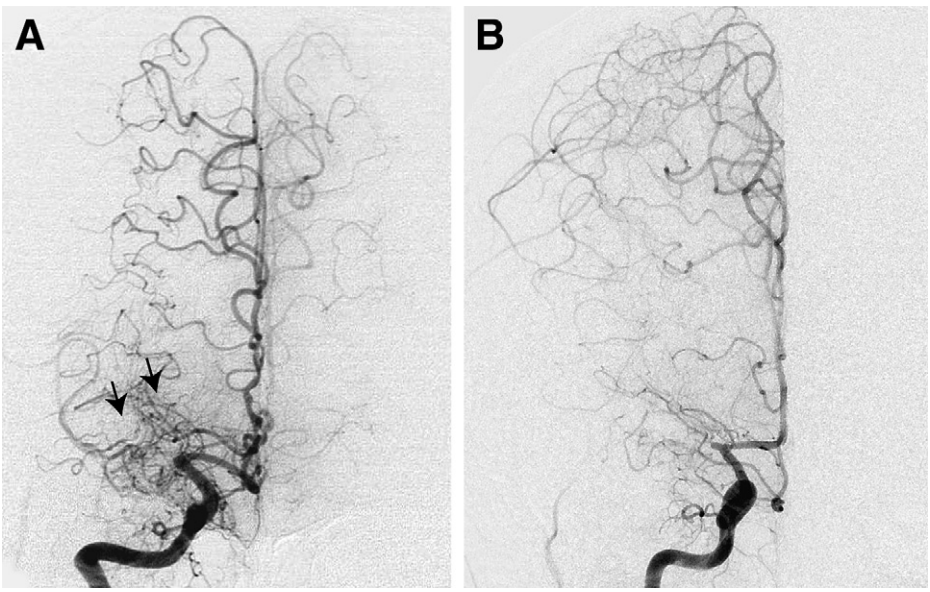

Figure 1. *Representative DSA images in patients with and without moyamoya vessels. A, Anteroposterior view image of a patient with moyamoya vessels (patient 12). The arrows indicate moyamoya vessels at the base of the brain. B, Anteroposterior view image of a patient with right MCA occlusion (patient 23) with no apparent moyamoya vessels at the base of the brain.*