Moyamoya Disease Progressing from Unilateral to Bilateral Involvement

—Case Report—

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Abstract

A 2-year-old boy with unilateral moyamoya disease demonstrated progression to the bilateral condition 1 year later. Previous reports of progression from unilateral to bilateral moyamoya disease showed that all patients were children or relatively young, and developed ischemic symptoms as the initial manifestation. These findings suggest that a considerable proportion of unilateral pediatric cases are in the initial stage of moyamoya disease and will progress to the contralateral side in the future.

Key words: moyamoya disease, basal ganglion, infarction

Introduction

The diagnostic criteria proposed by the Research Committee on Moyamoya Disease of the Ministry of Health and Welfare of Japan emphasize that the characteristic angiographic findings must be present bilaterally to diagnose a case as "definite."²⁾ Similar cases with unilateral involvement are classified as "probable." Although unilateral moyamoya disease is quite frequently reported, the relationship between the unilateral and bilateral entities is not clear. We report a case of unilateral moyamoya disease with later progression to the contralateral side, and discuss the relationship with definite bilateral moyamoya disease.

Case Report

A 2-year-old boy developed acute motor weakness in his right upper and lower extremities. He had experienced transient difficulty in walking, with headache and slight fever, 2 months previously. On admission, June 4, 1990, he was obtunded with right hemiparesis (including the face) and difficulty in speech. Precontrast computed tomographic (CT) scans disclosed a small infarction in the left inter-

nal capsule (Fig. 1 *left*). Postcontrast CT scans and magnetic resonance (MR) images found no moyamoya vessels in the basal ganglia, but poor visualization of the initial part of the left middle cerebral artery (MCA) (Fig. 1 *center, right*). Single photon emission CT scans with technetium-99m-hexamethyl propyleneamine oxime showed an extensive low-perfusion area in the left cerebral hemisphere.

He achieved gradual neurological recovery after admission and could walk unaided. Cerebral angiograms under general anesthesia on June 19 disclosed severe stenosis of the left internal carotid artery (ICA) at the terminal portion, involving the initial part of the left MCA (Fig. 2 *upper*). Lenticulostriate moyamoya vessels were unremarkable and no collateral circulation through transdural anastomosis was visualized. No distinct occlusive lesion was visible in the right ICA (Fig. 2 *lower*) or vertebrobasilar arteries. The diagnosis was unilateral moyamoya disease.

A left superficial temporal artery (STA)-MCA anastomosis with encephalo-myo-synangiosis (EMS) was performed on June 29. He was discharged on July 13, still with right hemiparesis and disturbance in speech. He did well for about 1 year, without recurrence of ischemic attacks.

He developed motor weakness of the left upper and lower extremities, contralateral to the previous

Fig. 1 *left*: Precontrast CT scan, showing a small low-density area in the left internal capsule. *center*, *right*: Postcontrast CT scan (*center*) and T₂-weighted MR image (*right*), showing poor demonstration of the initial part of the left MCA.

Fig. 2 *upper*: Left carotid angiograms, demonstrating severe stenosis at the terminal portion of the ICA. *lower*: Right carotid angiograms, showing no apparent occlusive lesion. *left*: Anteroposterior view, *right*: lateral view.

Fig. 3 *upper*: Right carotid angiograms, showing newly developed stenosis in the ICA. *lower*: Left carotid angiograms, showing progression of the stenosis in the ICA. *left*: Anteroposterior view, *right*: lateral view.

stroke, on July 11, 1991, and was readmitted. CT scans confirmed a newly developed cerebral infarction in the right centrum semiovale. Cerebral angiograms on August 16 showed a definite occlusive lesion at the terminal portion of the right ICA, which had appeared normal previously (Fig. 3 *upper*). Transdural anastomosis through the middle menin-

geal artery was also noted. The previous stenosis of the left ICA had also progressed (Fig. 3 *lower*). The left external carotid angiograms showed the STA-MCA bypass to be patent, with collateral circulation through the EMS and other external carotid vessels on the left.

An encephalo-duro-arterio-synangiosis was per-

formed on the right on August 26. Although he showed considerable neurological recovery, he remained tetraparetic at discharge on September 4.

Discussion

The relationship between unilateral and bilateral moyamoya disease is controversial.⁸⁾ Kitamura and Matsushima⁵⁾ found no apparent differences either in age distribution or in clinical manifestation among 23 unilateral cases compared to bilateral moyamoya disease. There were two peaks in age distribution in unilateral cases, one in childhood and the other in adult life. Most juvenile cases presented with cerebral ischemic symptoms as the initial manifestation, while hemorrhage was most common in adult patients.

Six previous cases have shown a progression from unilateral to bilateral lesions (Table 1).^{1,3,4,7,9)} Although the sample is too small to draw any conclusions, some unique features can be pointed out. First, five of seven cases, including ours, were children, and the two adult cases (>15 years old) were also relatively young, at 27 and 16 years old. Secondly, all patients except one without description developed ischemic symptoms, either transient ischemic attack or stroke, as the initial manifestation. These findings are easily understood in the context of previous documentation of angiographic progression of occlusive lesions in childhood moyamoya disease.⁶⁾ We suspect that a considerable proportion of unilateral pediatric cases are in the initial stage of moyamoya disease and will progress to bilateral disease in the future. In contrast, most adult cases, especially those with intracranial

hemorrhage at onset, are stable and remain unilateral.

Our case presented with cerebral infarction in the basal ganglia, which is rare in moyamoya disease,¹⁰⁾ since regional cerebral blood flow is generally preserved in this region by the abundant moyamoya vessels. Hayakawa *et al.*³⁾ also reported a pediatric case of moyamoya disease with cerebral infarction in the basal ganglia (Table 1). Their case resembles ours in age and later progression, indicating that when the moyamoya disease process is in the initial stage and the development of moyamoya vessels is not adequate, basal ganglionic infarction can occur due to inadequate blood flow.

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Table 1 Clinical summary of unilateral moyamoya disease with later progression to the contralateral side

Author (Year)	Age/Sex	Initial manifestation	Interval*
Mizukawa (1976) ⁹⁾	child/?	?	?
Hayakawa <i>et al.</i> (1989) ³⁾	1/M	lt carotid stroke	3 yrs 4 mos
Aoki <i>et al.</i> (1989) ¹⁾	27/F	rt carotid TIA	3 yrs 9 mos
Inoue <i>et al.</i> (1991) ⁴⁾	7/F	lt carotid TIA	2 yrs 9 mos
	8/F	rt carotid TIA	1 yr 10 mos
Kurose <i>et al.</i> (1991) ⁷⁾	16/F	lt carotid TIA	3 yrs
Present case	2/M	lt carotid stroke	1 yr

*From the initial angiographic diagnosis of unilateral moyamoya disease to that of bilateral moyamoya disease. TIA: transient ischemic attack.

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