Magnetic Resonance Angiography Demonstrating Adult Moyamoya Disease Progressing from Unilateral to Bilateral Involvement

—Case Report—

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Abstract

A 21-year-old woman presented with moyamoya disease manifesting as speech disturbance and right quadrant hemianopsia on October 22, 1994. Magnetic resonance (MR) angiography showed occlusion of the left internal carotid artery (ICA) with the normal right ICA. The diagnosis was “unilateral” moyamoya disease by conventional angiography. Follow-up MR angiography revealed further occlusive changes of the right middle cerebral artery (MCA) trunk on July 30, 1995, which progressed to occlusion of the MCA on March 25, 1997. Conventional angiography confirmed occlusion of the right terminal ICA to MCA with basal moyamoya vessels. The diagnosis was “bilateral” moyamoya disease. She was successfully treated by bilateral superficial temporal artery-MCA anastomosis. Follow-up MR angiography should be performed in relatively young patients with “unilateral” moyamoya disease to detect any progression to bilateral moyamoya disease.

Key words: moyamoya disease, magnetic resonance angiography, unilateral moyamoya disease

Introduction

Typical moyamoya disease is characterized angiographically by an abnormal network of moyamoya vessels in the “bilateral” base of the brain in association with progressive occlusion of the distal portions of the internal carotid arteries (ICAs). Moreover, the criteria established by the Japanese Ministry of Health and Welfare for the diagnosis of moyamoya disease require “bilateral” stenoocclusive changes verified by conventional angiography. “Unilateral” moyamoya disease, defined as “unilateral” typical angiographic evidence of moyamoya disease with contralateral normal cerebral vessels, may be an early stage of “bilateral” moyamoya disease or another clinical entity. Conventional angiography is the optimum method to investigate cerebral vessels, but is difficult to justify in patients with “unilateral” moyamoya disease. Magnetic resonance (MR) angiography provides an alternative to conventional angiography in patients with typical moyamoya disease. Therefore, follow-up studies with MR angiography could be important in clarifying the clinical entity of “unilateral” moyamoya disease.

We present an adult case of “unilateral” moyamoya disease with later progression to the contralateral side demonstrated by MR angiography and discuss the clinicoradiological features.

Case Report

A 21-year-old woman showed speech disturbance on October 22, 1994. Her past history was uneventful. Neurological examinations revealed motor and sensory speech disturbances in addition to dysgraphia and dyscalculia. Moreover, she had right lower quadrant hemianopsia. Computed tomography (CT) demonstrated a low density area (2 cm in diameter) in the left parietotemporooccipital region. MR imaging (Philips Gyroscan 1.5T; Philips Medical Systems, Amsterdam, Netherlands) revealed a low intensity area on the T1-weighted images and a high intensity
area on the T2-weighted images in the left parietotemporocipital region. MR angiography suggested occlusion of the left ICA with normal visualization of the right ICA and middle cerebral artery (MCA) (Fig. 1A). Conventional carotid angiography demonstrated left ICA occlusion at the C2 portion with abnormal vasculatures in the base of the brain, and normal right ICA and MCA (Fig. 2). Vertebral angiography demonstrated development of collaterals in the left cerebrum. The diagnosis was “unilateral” moyamoya disease. She was conservatively treated because her cerebral blood flow (CBF) had not decreased severely. Her speech disturbance completely recovered within a few weeks, and dysgraphia and dyscalculia improved gradually. She was followed up as an outpatient.

Follow-up MR angiography demonstrated severe stenosis of the right ICA to MCA on July 30, 1995 (Fig. 1B), but no new neurological deficits or ischemic lesions were observed on CT. Follow-up CT revealed a new ischemic lesion in the right frontoparietal region on March 25, 1997. MR angiography indicated occlusion of the bilateral ICAs (Fig. 1C). Conventional carotid angiography confirmed occlusion of the right ICA terminal portion with basal moyamoya vessels (Fig. 3). Single photon emission CT revealed low perfusion areas in the left parietotemporocipital and the right frontoparietal regions. According to our criteria for direct revascularization in adult moyamoya disease,16) right superficial temporal artery (STA) to MCA anastomosis was performed on July 15, 1997 by double anastomoses, in which the bypass flow was 55 ml/min measured by an electromagnetic flowmeter (MFV 1100; Nihon Kohden Corp., Tokyo) and the cortical artery pressure was 26 mmHg as defined as the back pressure measured during temporary occlusion of the STA trunk upon completion.
Fig. 3 Follow-up carotid angiograms demonstrating right internal carotid artery occlusion at the terminal portion with abnormal vessels in the base of the brain (A: anteroposterior view, B: lateral view).

of the anastomosis. Left STA-MCA anastomosis was performed on December 3, 1997 by single anastomosis, in which the bypass flow was 30 ml/min and the cortical artery pressure was 28 mmHg. These intraoperative hemodynamic data were consistent with those of typical moyamoya disease.16) MR angiography and CBF studies demonstrated improvement of cerebral circulation via the STA. Her postoperative course was good.

Discussion

MR angiography demonstrated the progression of “unilateral” adult moyamoya disease to typical “bilateral” moyamoya disease in the present case. The term “unilateral” moyamoya disease is used to describe angiographical evidence of stenosis or occlusion of a major intracranial artery in association with abnormal fine vessels on one side.4,9,20) The diagnostic criteria of the Research Committee on Moyamoya Disease of the Ministry of Health and Welfare, Japan emphasize that the characteristic angiographic findings must be bilateral.14) However, some moyamoya disease patients have mild unilateral or bilateral arterial stenosis at the beginning of the disease and progress to a definite lesion.1,2,5,18) Therefore, the “unilateral” lesion may represent the early stages of a “bilateral” lesion. However, only a small number of cases with progressive involvement from “unilateral” to “bilateral” moyamoya disease are known.6,8,11,12,19,22)

“Unilateral” moyamoya disease may progress to “bilateral” moyamoya disease as angiographic changes are initiated in the unaffected vessels. Analysis of six cases that demonstrated progression from “unilateral” to “bilateral” moyamoya disease found that “unilateral” pediatric cases are in the initial stage of moyamoya disease and later progress to “bilateral” disease.22) Among 32 patients with “unilateral” occlusive carotid disease who received angiographic follow-up examinations, 17 patients, 12 children and five adults, developed “bilateral” lesions between 1 and 7 years after the diagnosis of “unilateral” lesion.10) Most of these patients demonstrated ischemic attacks as the initial episode and had repeated attacks before admission. Young children, mostly less than 10 years of age, tended to develop “bilateral” lesions within 1–2 years, but adults tended to demonstrate only a “unilateral” lesion. Therefore, children or young adults with “unilateral” occlusive lesions of the terminal portion of the ICA are likely to develop “bilateral” disease within 1–5 years.

Longitudinal angiographic changes, familial occurrence, and the basic fibroblast growth factor concentration were analyzed in the cerebrospinal fluid of patients with “unilateral” moyamoya disease.6) Over a 10-year period, 10 patients with “unilateral” moyamoya disease were followed up using conventional angiography or MR angiography. Only one pediatric patient demonstrated obvious signs of progression to typical bilateral disease. There was no familial occurrence among these 10 cases, and the levels of basic fibroblast growth factor concentration were low. These findings suggest that most cases of “unilateral” moyamoya disease, especially in adults, are distinct from typical “bilateral” moyamoya disease. Only 13 adult (over 16 years old) cases, five females, three males, and five unreported, have demonstrated progression from “unilateral” to “bilateral” moyamoya disease.3,8,12,15,22) In these 13 cases and ours, some distinctive features were observed, such as relatively young age, 16–63 years old (mean 34 years), initial symptom of ischemic stroke in all cases except one had hemorrhagic attack at 63 years of age, and progression from “unilateral” to “bilateral” lesions in eight of the 14 cases occurred in less than 3 years. These findings suggest that “unilateral” adult moyamoya disease should be closely followed up for at least 3 years after the initial attack.

Combined application of MR imaging and MR angiography are useful for detecting moyamoya disease.15) More than 80% of suspected moyamoya disease cases can be correctly diagnosed by MR angiography.21) Analysis of 39 moyamoya disease patients concluded that MR angiography is an alternative to conventional angiography in typical moyamaya disease.7) MR angiography has demonstrated some problems in the definitive diagnosis of
moyamoya disease due to the different angiographic features relevant to the stages, such as atherosclerotic cerebral arterial occlusion in the end stage of adult moyamoya disease. However, MR angiography is less invasive than conventional angiography, and can be repeated many times, so is a better follow-up method for moyamoya disease. In our case, progressive occlusive changes of the ICA to MCA were observed at the early stage when no new neurological deficits or ischemic lesions were observed on CT.

This rare case of “unilateral” adult moyamoya disease, which was confirmed to progress to “bilateral” involvement by MR angiography, suggests that relatively young adult patients with “unilateral” moyamoya disease should be closely followed by MR angiography for at least 3 years after the initial attack.

References