Adult Moyamoya Disease Progressing from Unilateral to Bilateral Involvement

—Two Case Reports—

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Abstract

A 30-year-old female and a 38-year-old male developed “definite” moyamoya disease after presenting with “probable” moyamoya disease manifesting as ischemic stroke. Angiography at first presentation demonstrated unilateral involvement with normal contralateral findings. The cases were classified as adult type “probable” moyamoya disease according to the diagnostic criteria. Follow-up angiography demonstrated the occlusive lesions had become bilateral, satisfying the criteria for “definite” cases. Serial angiography is required in adults presenting with “probable” moyamoya disease manifesting as ischemic stroke.

Key words: moyamoya disease, adult, unilateral moyamoya disease, “probable” moyamoya disease

Introduction

Moyamoya disease is characterized by angiographic findings of bilateral stenoses or occlusions at the terminal portion of the internal carotid artery (ICA) and the proximal anterior (ACA) and middle cerebral arteries (MCA), together with abnormal vascular networks (so-called moyamoya vessels) in the arterial phase near the stenosis or obstruction. The diagnostic criteria of the Japan Cooperative Research Committee (JCRC) on occlusion of the circle of Willis classify adult cases with bilateral occlusive lesions as “definite” moyamoya disease (bilateral moyamoya disease) and those with unilateral involvement as “probable” moyamoya disease (unilateral moyamoya disease). It is unclear whether both the adult “definite” and “probable” moyamoya disease types belong to the same entity.

Adult “probable” moyamoya disease cases which progress from unilateral to bilateral involvement are extremely rare. We describe two such cases demonstrating rapid development from unilateral to bilateral involvement.

Case Reports

Case 1: A 30-year-old female described previously first suffered transient ischemic attack (TIA) on the left side in October 1981. After the first attack, she had repeated episodes of transient left hemiparesis. On admission, neurological examination demonstrated normal findings except for hyperactive tendon reflexes in her left extremities. Computed tomography (CT) showed no abnormality. Right carotid angiography revealed stenosis of the ICA at the terminal portion and moyamoya vessels near the terminal portion of the ACA, and
moyamoya vessels near the stenoses (Fig. 2 upper row). Collateral circulation from the vertebrobasilar system supplied the bilateral ICA territories. Moreover, right carotid angiography showed the stenosis of the ICA at the terminal portion, the occlusion of the ACA, and faint basal moyamoya vessels (Fig. 2 lower row). Xe-enhanced CT revealed decreased cerebral blood flow in the right hemisphere. A modified encephalo-duro-arterio-synangiosis (EDAS) was performed on the right side. The TIA episodes decreased in frequency after surgery and she is now doing well without TIA symptoms.

Case 2: A 38-year-old male suffered from slowly progressive right hemiparesis in 1986. Neurological examination on admission in May 1986 demonstrat-
ed mild right hemiparesis. CT disclosed an infarction in the left frontal cortical area. Left carotid angiography showed occlusion of the ICA at the terminal portion and moyamoya vessels near the occlusion. Right carotid and vertebral angiography demonstrated no occlusive lesions or moyamoya networks. No obvious basic disorder related to cerebrovascular occlusion or stenosis was detected. EDAS was performed on the left side to treat the "probable" moyamoya disease.

Postoperative left carotid angiography 6 months after surgery showed the occlusion of the ICA at the terminal portion and poor collateral circulation from the EDAS (Fig. 3 left). Right carotid angiography showed no abnormal findings (Fig. 3 center, right). No ischemic or hemorrhagic episode occurred for 2 years, but he then suddenly developed severe headache and generalized convulsion, and afterwards rapidly became comatose.

CT demonstrated a massive cerebral hematoma in the right frontotemporal lobes with intraventricular hematoma. Left carotid angiography on the same day showed no changes compared with the previous study, but right carotid angiography disclosed the stenosis of the ICA at the terminal portion and moyamoya vessels near the stenosis (Fig. 4). He did not recover from the sequelae of intracerebral hematoma and died 3 days later. Autopsy was not permitted.

**Discussion**

The controversy about the diagnosis of adult type "probable" moyamoya disease centers on whether the so-called moyamoya vessels might be a manifestation of another systemic disease, particularly cerebral arterial occlusion due to arteriosclerosis or rete mirabile.5)

Kitamura and Matsushima8) reported no apparent difference in age distribution or clinical manifestation between "probable" and "definite" moyamoya disease patients. They also reported that five of 11 cases with adult "probable" moyamoya disease had occlusive lesions of the ICA or ACA and moyamoya vessels on the opposite side. These lesions are outside the diagnostic criteria of "definite" moyamoya disease. Kitamura7) also classified cases of "probable" moyamoya disease into three types according to the findings on the side opposite to the occlusive changes: 1) very minimal stenosis, 2) atypical findings, and 3) no abnormalities. Our cases belong to the third group.

We previously reported that 11 of 17 patients with adult type "probable" moyamoya disease had angiographic evidence of occlusion of the extracranial ICA, posterior cerebral artery, or ACA (4 cases) and/or leptomeningeal anastomosis or transdural anastomosis (9) on the opposite side.14) The JCRC diagnostic criteria for "definite" moyamoya disease exclude these lesions, but these findings strongly suggest that adult type "probable" moyamoya disease is akin to "definite" moyamoya disease. Previous and present reported cases have included nine demonstrating development of unilateral to bilateral moyamoya disease.1,3,4,9-12,17) Six cases developed in childhood with ischemic stroke and three in people aged 16 years or older. Despite the small number, some distinctive features can be seen. The adult cases were relatively young, at 16, 27, and 37 years old. The initial symptom was ischemic stroke, whereas adult cases with "definite" moyamoya disease often have hemorrhagic stroke as the initial manifestation and are stable angiographically.

These cases strongly suggest that "probable" moyamoya disease with ischemic stroke as the initial manifestation may develop into "definite" moyamoya disease. Therefore, careful attention should be paid to symptomatic changes and serial angiography in adults with "probable" moyamoya disease presenting with ischemic stroke.

**References**


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