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Case Report

Rapid Progression of Unilateral Moyamoya Disease

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The detailed clinical characteristics of unilateral moyamoya disease (MMD) have not been fully elucidated. It has been reported that some patients with unilateral MMD progress to bilateral involvement, while others remain with the unilateral variant. In this series, we present a case of unilateral MMD that progressed to bilateral involvement over the course of just one month.

Key Words : Unilateral moyamoya disease · Progression.

INTRODUCTION

Moyamoya disease (MMD) is characterized by unknown etiology with progressive bilateral stenosis or occlusion of the internal carotid artery (ICA) with the formation of a vascular network, the so called 'moyamoya vessels'. When these angiographic findings are unilateral, the case is considered probable MMD.

Young children with unilateral MMD tend to develop bilateral lesions, but the lesion tends to remain unilateral in adults. Rapid progression of unilateral MMD to bilateral MMD has most frequently been reported in infants.[2-6,14] In this study, we report a case of rapid progression from unilateral MMD to bilateral involvement, and prudently assume that definite MMD results from unilateral MMD.

CASE REPORT

A 13-month-old boy presented with right hemiparesis (grade I/I) and rigidity. He was born at gestational age 30+2 weeks by vaginal delivery. Both parents were healthy and had no history of congenital disease.

Brain computed tomography (CT) revealed a diffuse hypodense lesion in the territory of the left middle cerebral artery (MCA) and brain magnetic resonance (MR) diffusion-weighted imaging revealed diffusion restriction in the left MCA territory (Fig. 1). Cerebral angiography showed severe, diffuse stenosis involving the supraclinoid segment of the left ICA and proximal M1 segment of the left MCA, while the right carotid and posterior circulations appeared normal (Fig. 2). We were hesitant to perform surgical procedures because infants are presumed to be vulnerable to perioperative complications. In two weeks after ischemic attack, the motor strength in the left arm and leg was improved to grade II/III and the patient was discharged.

One month later, he presented with left hemiparesis (grade I/I) and seizure. Brain MR diffusion-weighted imaging revealed diffusion restriction of the right MCA territory (Fig. 3), and magnetic resonance angiography showed occlusion of the right distal

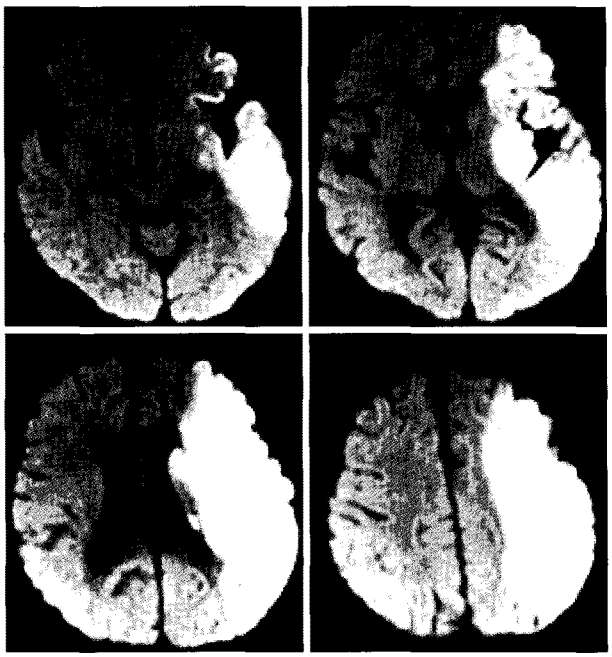

Fig. 1. Brain diffusion magnetic resonance image showing diffusion restriction of the middle cerebral artery territory consistent with the cerebral infarction.

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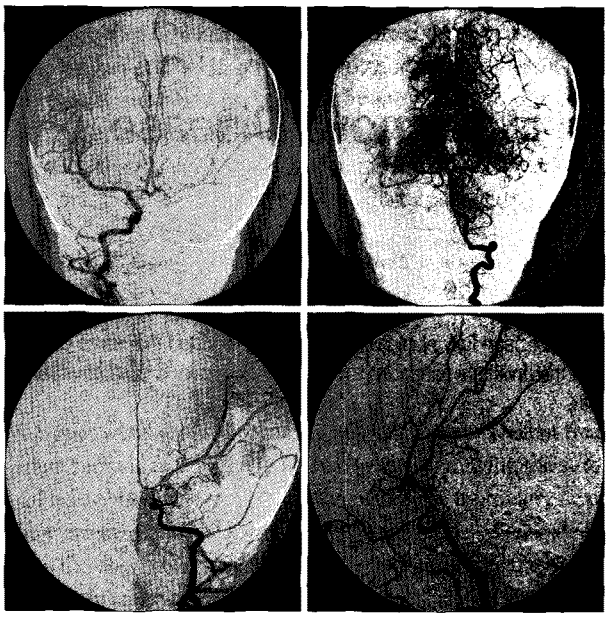

Fig. 2. Cerebral angiography of the left internal carotid artery (ICA) revealing severe, diffuse stenosis involving the supraclinoid segment of the left ICA and proximal left middle cerebral artery (MCA). However, other angiographic studies showed normal appearance of right carotid and posterior circulation, as well as collateral flow in the left MCA via the anterior communicating artery.

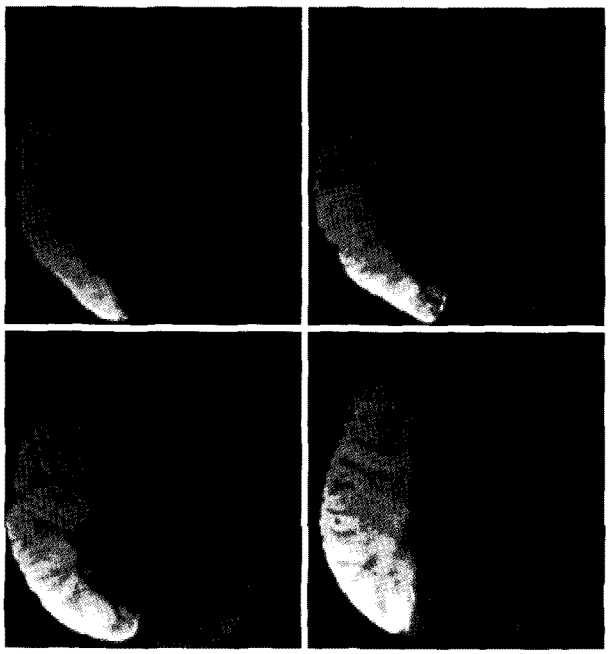

Fig. 3. Brain diffusion magnetic resonance image taken at the time of the second attack showing diffusion restriction of the right middle cerebral artery territory.

ICA and severe stenosis of the right posterior cerebral artery (Fig. 4). Several studies, including chromosome studies, were performed, but there was no clinical evidence of moyamoya syndrome. The patient was progressed to vegetative state and died in seven months after initial attack.

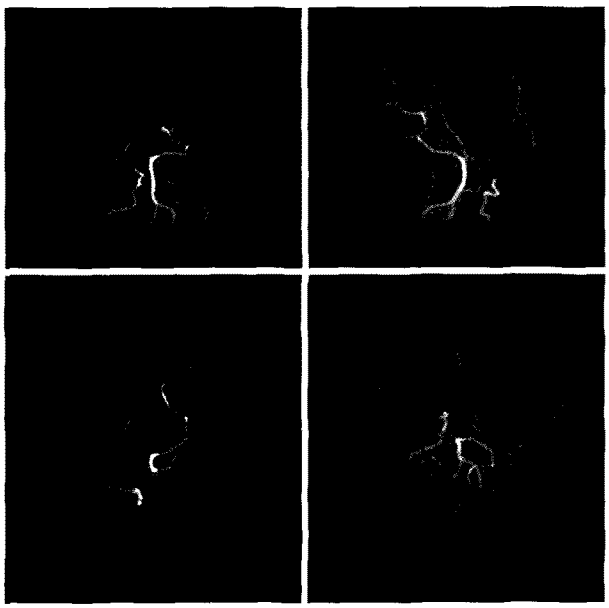

Fig. 4. Brain magnetic resonance angiography showing the occlusion of the distal portion of the right internal carotid artery and severe stenosis of the right posterior cerebral artery.

DISCUSSION

Multiple infarctions in the developing brain, especially those that affect both hemispheres, lead to poor clinical outcomes, intellectual impairment and developmental delays in patients with MMD.[12] Therefore, early diagnosis and treatment are crucial for avoiding irreversible neurologic deficits. Matsushima et al.[9] recommended that patients under 2 years of age with unilateral MMD should undergo a single bilateral operation because of the high frequency of subsequent lesion development on the normal side.

Patients with unilateral MMD, especially children, often exhibit progression to typical bilateral MMD.[3,5,6,9] We reviewed the literature regarding the time and prevalence of progression from unilateral MMD to bilateral MMD and the methods used to treat this lesion (Table 1).[1-8,10-14] The duration of progression of unilateral MMD to bilateral MMD varies. Hirotsune et al.[2] reported a mean time of 20 months (range 4-34 months), and Houkin et al.[3] reported only one pediatric case of bilateral MMD among 10 cases of unilateral MMD evaluated at a 6-month follow-up examination. Kawano et al.[5] reported that bilateral lesions are likely to develop within 1 to 2 years in young children with unilateral evidence of MMD. To date, the most rapid time to progression was 4 months. Approximately ten previous cases showed progression times within 1 year. These rapid progression findings suggest that a considerable proportion of pediatric cases of unilateral MMD are in the initial stage of MMD and will eventually progress to bilateral involvement.[14] In this report, we experienced a case of rapid progression at the one-month follow-up. We assume that MMD does not necessarily develop bilaterally in a synchronized fashion but that unilateral MMD is an early form

Table 1. Literature review of reported cases of progressive involvement from unilateral MMD to bilateral MMD

Authors & years	Case no. of unilateral MMD	Age	Prevalence of bilateral progression	Time to progression	Treatment for bilateral MMD
Matsushima, 1994	6	6, 8	33.3% (2/6)	2.16 years (1.66-2.66)	Indirect revascularization in 2 cases
Kawano, 1994	64	14.1 (0-63)	26.5% (17/64)	2.56 years (0.8-7)**	
Houkin, 1996	10	4	10% (1/10)	0.5 year*	
Hirotsune, 1997	17	6.16	41% (6/17)	20 months (4-34)**	Indirect revascularization in 6 cases
					Combined (direct+indirect) in 1 case
Seol, 2006	7	5, 8	28.5% (2/7)	29 months (27 & 32)	Indirect revascularization
Kelly, 2006	18	32.4	39% (7/18)	12.7 months (5-22)***	Direct revascularization in 4 cases
Smith, 2008†	33		30% (10/33)	2.2 years (0.5-8.5)	
				0.9 years (age <7)†	
				3.1 years (age ≥7)	
Murphy, 1980‡	1	29		2 years	Direct revascularization
Kurose, 1991‡	1	16		3 years	Direct revascularization
Yoshida, 1992‡	1	2		1 year*	Combined (direct+indirect)
Wanifuchi, 1996‡	2	30, 38		4 years, 2 years	Indirect revascularization
Fujiwara, 1997‡	1	54		49 months	Direct revascularization
Kagawa, 2004‡	1	21		9 months*	Direct revascularization

*One case of progression within 1 year, **Two cases of Progression within 1 year, ***Three cases of progression within 1 year, †Smith et al did not report the summary of cases of progression. Therefore we could not know the age and treatment method, ‡Case reports. MMD : moyamoya disease

of definite MMD.

CONCLUSION

To date, several cases of MMD with progression from unilateral to bilateral involvement have been reported. The time to progression varied from 4 to 49 months. This case illustrates very rapid progression of MMD, being only one month after initial presentation. It is reasonable to assume that unilateral MMD in infants is the early phase of the pathologic process of definite MMD.

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