Moyamoya syndrome in young children: MR comparison with adult onset.

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Moyamoya Syndrome in Young Children: MR Comparison with Adult Onset

Jun-ichi Takanashi,[1] Katsuo Sugita,[1] Mitsuko Ishii,[1] Yuzo Tanabe,[2] Chiaki Ito,[3] Hiroaki Date,[3] and Hiroo Niimi[1]

PURPOSE: To clarify whether there were any differences in MR appearance between the childhood and the adult moyamoya syndromes. **METHOD:** We compared the cranial MR findings in four children under the age of 6 who had moyamoya syndrome with previously documented adult cases. **RESULTS:** Moyamoya syndrome in younger children exhibited a significant increase in cortical and subcortical infarction, and a decreased incidence of deep white matter infarction in the centrum semiovale and basal ganglia, in contrast to adult cases. There were no remarkable differences between these two groups of moyamoya cases with regard to the occlusive changes of the internal carotid and middle cerebral arteries, or to the flow void sign on MR. **CONCLUSION:** These differences in the sites and frequencies of infarctions between the childhood and the adult moyamoya syndromes observed on MR might reflect differences in the cerebral circulation.

Index terms: Moyamoya disease; Arteries, carotid; Blood vessels, flow dynamics; Brain, magnetic resonance; Pediatric neuroradiology

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Moyamoya syndrome is a rare cerebrovascular occlusive disease characterized by the angiographic appearance of an abnormal vascular network at the base of the brain, known as moyamoya vessels (1–4). Angiography is not always safe, especially in children (5), although it is essential for a diagnosis. Magnetic resonance (MR) has been reported recently to be valuable in the diagnosis of moyamoya syndrome in allowing the detection of occlusive changes of internal carotid artery (ICA) and middle cerebral artery (MCA), moyamoya vessels in the basal ganglia and thalamus, and cortical and subcortical infarction (6–10). The reported ages when MR was performed of patients with moyamoya syndrome have ranged from 17 to 40 years (mean age, 34 years) (6), 6 to 43 years (mean age, 16.3 years) (7), and 6 to 33 years (8). These reports, however, did not mention any differences with age, although the clinical manifestations are remarkably different between the childhood and adult moyamoya syndromes. The former usually comprise transient cerebral ischemic attacks and infarctions, and the latter hemorrhage in the brain (5, 11). We, therefore, examined the MR findings in moyamoya syndrome patients under the age of 6, and compared them with those in adult cases described previously.

Patients and Methods

Four patients (two boys and two girls, Table 1), 1 to 6 years old, were diagnosed as having moyamoya syndrome from the typical angiographic appearance. According to the angiographic classification of Suzuki (stage 1 is stenosis in the region of distal ICA bifurcations only; stage 2, appearance of moyamoya; stage 3, growth of moyamoya and disappearance of anterior cerebral arteries [ACA] and MCA; stage 4, decrease in moyamoya and disappearance of posterior cerebral arteries; stage 5, further decrease in moyamoya and disappearance of all the major cerebral arteries; and stage 6, disappearance of moyamoya and blood supply only from the external carotid arteries) (3), one hemisphere was classified as stage 2, three as stage 3, two as stage 4, and two as stage 5 (Table 2). The clinical manifestations at the onset were monoplegia in three cases (cases 1, 2, and 3) and hemiconvulsions in the other (case 4).

Cranial MR scanning was performed with a 0.5-T superconducting magnet (General Electric, Milwaukee, WI) in

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[1] Department of Pediatrics, School of Medicine, Chiba University, 1-8-1 Inohana, Chuo-ku, Chiba-shi 260, Japan. Address reprint requests to J. Takanashi, MD.

Divisions of [2]Neurology and [3]Neurosurgery, Chiba Children's Hospital, 579-1 Heta, Midori-ku, Chiba-shi 266, Japan.

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three cases (cases 1, 2, and 3), and a 1.5-T superconducting magnet (General Electric) in the other (case 4). The imaging parameters for the 0.5-T unit were as follows: matrix size, 224 × 160; field of view, 25 cm; and section thickness, 7 mm. Axial and coronal T2-weighted images with a spin-echo pulse sequence (2000/80/2, repetition time/echo time/excitations) and T1-weighted images with an inversion-recovery pulse sequence (2000/25/2, inversion time = 700) were obtained in all three cases. The parameters for the 1.5-T unit were as follows: matrix size, 256 × 192; field of view, 20 cm; and section thickness, 5 mm. Axial and coronal T2-weighted (3000/100/1) and proton-density–weighted (3000/30) images were obtained. The MR images were reviewed with regard to arterial occlusive changes, collateral pathways, and infarctions. The cavernous and supraclinoid ICA was defined as being narrowed when the diameter was definitely smaller than that of the basilar artery or the contralateral ICA. MCA was taken as being occluded when signal void caused by flow was not detected. Cranial MR was performed within 2 weeks after the onset of symptoms in two cases (cases 3 and 4) and after an interval of 2 to 3 years in the other two cases (cases 1 and 2).

Results

Infarction

The parenchymal abnormalities observed are summarized in Table 2. Cerebral infarctions were diagnosed as abnormally low signal areas on T1-weighted images, with corresponding abnormally high signal areas on T2-weighted images. Cortical and subcortical infarctions were observed in all four patients, seven out of eight hemispheres (Figs. 1, A and B, and 2, C and D), being located not only in the watershed regions between the MCA and the ACA and between the MCA and the posterior cerebral artery, but also within the territories supplied by the ACA and the MCA. Infarction in the basal ganglia, thalamus, and internal capsules was not detected in any case. The centrum semiovale was affected in three patients, three out of eight hemispheres (two at stage 4 and one at stage 5; Fig. 1C).

Vascular Abnormalities

Narrowing of the ICA was observed in two out of eight carotid arteries (cases 2 and 4, angiographic stages 3 and 4, respectively; Fig. 2A). Narrowing or occlusion of the MCA was detected in seven of eight. Moyamoya vessels, appearing as multiple, small, round, or tortuous signal void areas in the basal ganglia and/or thalamus, were well observed on six sides (Fig. 1, A and B), but could not be detected on the other two (case 4, Fig. 2, B and C).

TABLE 1: Case presentations

Case	Age at Onset (yr)	Sex	Age at Scanning (yr)	Clinical Manifestations
1	3	F	6	Right hemiparesis
2	4	F	6	Leg weakness
3	5	M	5	Right leg monoparesis
4	1	M	1	Right hemiparesis, generalized tonic-clonic seizures

Discussion

Cortical and subcortical infarctions have been reported to occur in 30% to 42% of adult patients with moyamoya and to be located predominantly in hemodynamically vulnerable regions (6, 7) (ie, in the watershed regions between the ACA, MCA, and posterior cerebral artery). In our childhood

TABLE 2: Angiographic and MR imaging findings

Case	Angiographic Stage (rt/lt)	MR Imaging Findings							
		Occlusive Change		Moyamoya vessels	Infarction				
		ICA	MCA		CS	BG/Th	A-M	M-P	MCA
1	4	−	+	+	+	−	−	+	+
	3	−	+	+	−	−	−	−	+
2	3	−	+	+	+	−	−	+	−
	4	+	+	+	−	−	−	+	−
3	5	−	+	+	+	−	+	+	+
	5	−	+	+	−	−	+	−	+
4	2	−	−	−	−	−	−	−	−
	3	+	+	−	−	−	+	−	+

Note. rt indicates right; lt, left; CS, centrum semiovale; BG/Th, basal ganglia and thalamus; A-M, watershed infarction between ACA and MCA; M-P, watershed infarction between MCA and posterior cerebral artery; and MCA, infarction in the territory of MCA.

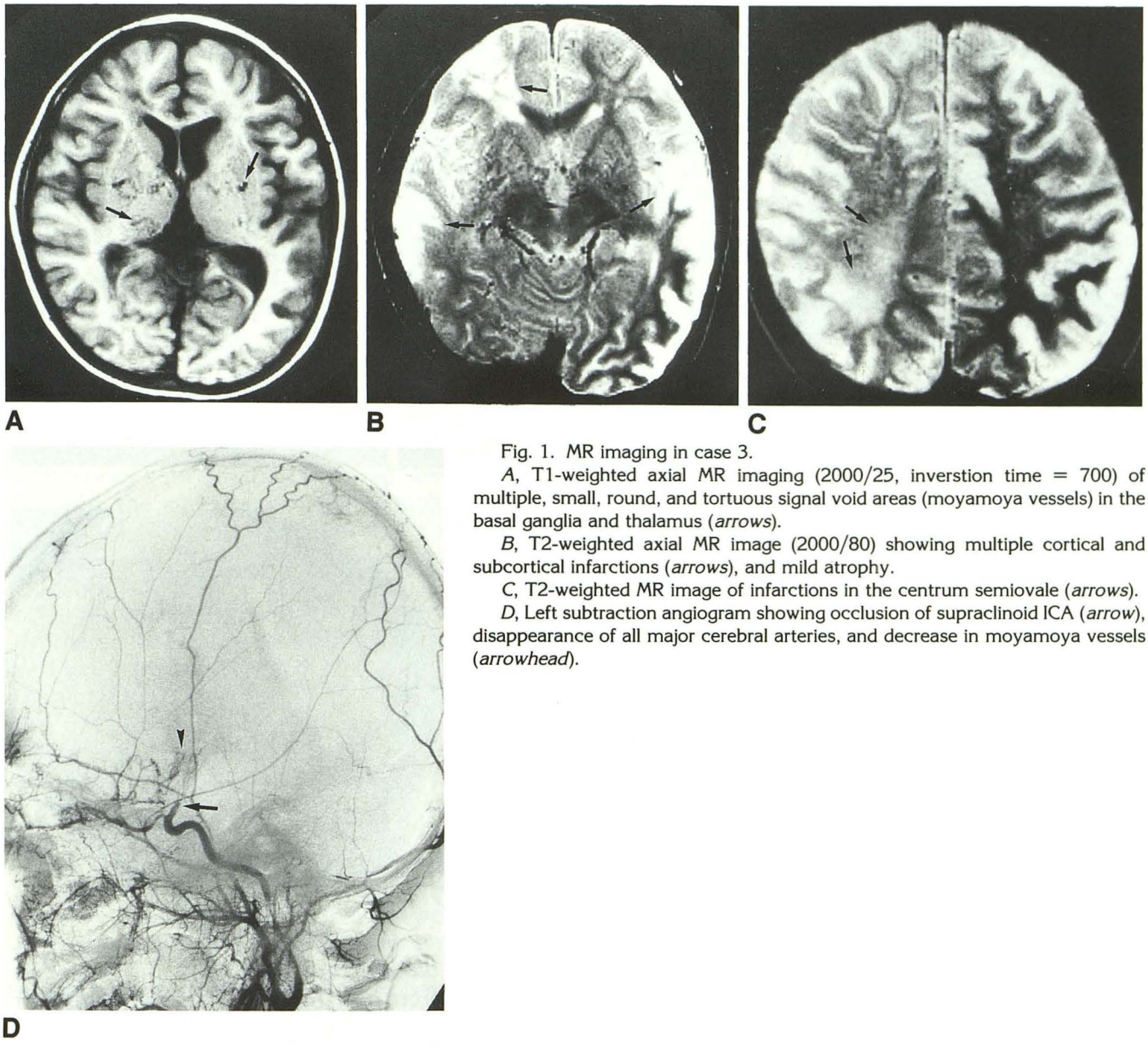

Fig. 1. MR imaging in case 3.

A, T1-weighted axial MR imaging (2000/25, inverstion time = 700) of multiple, small, round, and tortuous signal void areas (moyamoya vessels) in the basal ganglia and thalamus (*arrows*).

B, T2-weighted axial MR image (2000/80) showing multiple cortical and subcortical infarctions (*arrows*), and mild atrophy.

C, T2-weighted MR image of infarctions in the centrum semiovale (*arrows*).

D, Left subtraction angiogram showing occlusion of supraclinoid ICA (*arrow*), disappearance of all major cerebral arteries, and decrease in moyamoya vessels (*arrowhead*).

series, such infarctions were detected in all four patients, seven out of eight hemispheres, being located not only in watershed regions, but also within the territories supplied by MCA. In the light of cerebral blood flow (CBF), the differences in hemispheric CBF, as measured by single-photon emission computed tomography, between patients with moyamoya syndrome and control subjects, were more marked in the younger age group (12, 13). With advancing age, the differences decreased and eventually disappeared. Over the age of 40, no difference was detected in hemispheric CBF.

According to angiographic findings of cortical arteries in moyamoya syndrome (14), the occlusion of cortical arteries in adult cases is milder than that in childhood. It is, therefore, speculated that the high frequency of cortical or subcortical infarctions and their distribution in early childhood moyamoya syndrome probably reflect the decrease in cortical blood flow in addition to that in hemispheric CBF itself, both of which are characteristic in childhood.

From the viewpoints of clinical manifestations and courses, irreversible neurologic deficits and intellectual regression are more likely to develop in moyamoya syndrome under the age of 4 years (15). According to a report on intelligence quotients in moyamoya syndrome (16), the mean intelligence quotient of the younger group, the

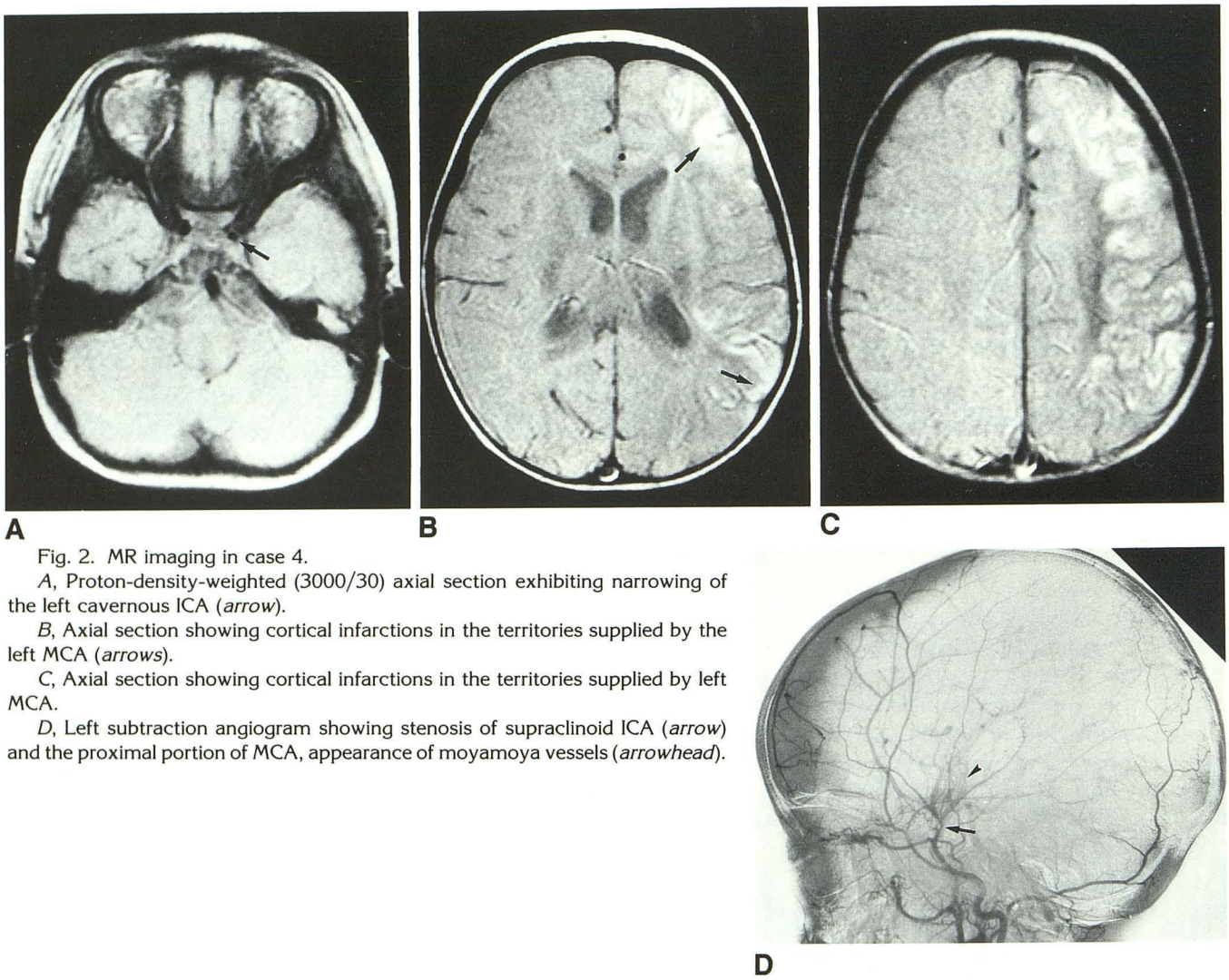

Fig. 2. MR imaging in case 4.

A, Proton-density-weighted (3000/30) axial section exhibiting narrowing of the left cavernous ICA (*arrow*).

B, Axial section showing cortical infarctions in the territories supplied by the left MCA (*arrows*).

C, Axial section showing cortical infarctions in the territories supplied by left MCA.

D, Left subtraction angiogram showing stenosis of supraclinoid ICA (*arrow*) and the proximal portion of MCA, appearance of moyamoya vessels (*arrowhead*).

age of onset in whom was under 6 years, was 87, significantly lower than that of the older group (intelligence quotient 123). These facts might also be explained by the clinical evidence that cortical and subcortical infarctions occur more frequently in childhood moyamoya syndrome.

On the other hand, the centrum semiovale is one of the most affected areas in adult moyamoya syndrome. For example, Bruno et al (6) reported that infarctions in the centrum semiovale were detected in all 12 hemispheres, and 56 (82%) out of 68 infarctions were observed at this site. Infarctions in the centrum semiovale have also been frequently observed in other carotid artery occlusive disorders. These lesions were believed to reflect the fact that the centrum semiovale is a hemodynamically vulnerable region supplied by long, penetrating branches of ACA and MCA. Interestingly, infarction in the centrum semiovale was observed in only three hemispheres (stages 4 and 5) out of the eight in our childhood series. Furthermore, no infarction was observed in the basal ganglia or thalamus, in contrast to in adult moyamoya syndrome, in which infarctions are detected at a rate of 39% to 50% (6, 7).

These differences might be explained by the age-associated change in the capacity to form collateral pathways between basal perforating vessels and medullary arteries arising from ACA and MCA. According to Kodama and Suzuki (17) and Takahashi (18), lenticulostriate arteries (some of the most important vessels among basal moyamoya vessels) become smaller in number and size with advancing age. The diameter of anastomoses between lenticulostriate arteries and medullary arteries decreases from 30 μm at 6 years to 20 μm at 32 years. They concluded that moyamoya vessels in younger cases are more

easily formed than in older ones. Moreover, as shown by the single-photon emission computed tomographic studies on regional CBF in moyamoya syndrome by Ogawa et al (13, 19), CBF in the parietal lobes of patients under the age of 9 is higher than that in the frontal and temporal lobes, but there are no differences in CBF in the frontal, temporal, and parietal lobes in patients over age 10. Thus, it is believed that infarctions are prevented in the deep white matter and basal ganglia in early childhood moyamoya syndrome, even at a continually ischemic stage, because of this excellent collateral circulation (moyamoya vessels).

Concerning the MR findings of both occlusive changes of ICA and MCA, and the flow void sign of moyamoya vessels, there were no remarkable differences between the childhood cases we studied and the one reported previously. The frequency of ICA stenosis in our cases (two out of eight vessels) was lower than those reported previously in adults (75% to 87%) (6, 7). The stenosis observed on MR, for example, was probably overestimated in Chang et al's (7) study in which they defined "narrowing" of ICA as a diameter smaller than or equal to that of the basilar artery. Therefore, we conceived that these frequencies could not be simply compared, because the criteria for stenosis of ICA were not clearly defined.

On the other hand, the flow void sign of moyamoya vessels was observed on MR at the rate of 82% to 96% (7, 8). In our series, moyamoya vessels were detected in six out of eight hemispheres. The angiographic stages were 2 and 3 in the right and left hemispheres of case 4, which exhibited no flow void sign. Hasuo et al (20) mentioned that the diagnosis of moyamoya syndrome on the basis of MR posed no problems at stages 3 and 4. Brady et al (9) reported a case with moyamoya syndrome in whom the flow void sign of moyamoya vessels was not distinctly recognized on MR until 10 months after the first scan, which revealed only periventricular infarctions. As a method for the screening for moyamoya disease, MR is useful even in early childhood. It should, however, be taken into consideration that occlusive changes of ICA and MCA and the flow void sign of moyamoya vessels cannot always be detected, at least before stage 3.

In conclusion, moyamoya syndrome in younger children exhibits a significant increase in cortical and subcortical infarction, and a decreased incidence of deep white matter infarction in the centrum semiovale and in the basal ganglia, compared with adult patients; second, the flow void sign of moyamoya vessels within the basal ganglia is not seen before stage 3 disease.

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