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Adult-onset Moyamoya Disease : Clinical Features and Prognosis

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Background : The age distribution of patients with moyamoya disease (MMD) forms two characteristic peaks in children and adults. It is well known that hemorrhagic presentation is more frequent in adults, while hemorrhage is rare in children. There is controversy in the management of adult-onset MMD and its natural course is not well defined. We investigated the clinical features and outcomes of adult onset MMD to clarify its characteristics. **Methods** : Fifty patients with MMD whose first manifestation appeared later than 15 years of age were studied retrospectively. The first manifestations were divided into ischemic and hemorrhagic categories. The types of reattack and their frequencies according to treatment type, as well as the differences between probable (unilateral) and definite (bilateral) MMD, and current clinical outcomes by the Modified Rankin Scale were investigated. **Results** : The mean follow-up duration was 24.4±25.9 months. The female/male ratio was 1.5. The most frequent age of onset was in the 4th decade. Ischemia was more frequent (58%) than hemorrhage (40%) at the initial presentation. Reattack occurred in 20 (41%) patients and more frequently in the ischemic type (55%) than the hemorrhagic (20%). Patients with probable MMD had a later onset age (39 vs. 31.5 years old) and more frequent reattacks (50%) than patients with definite MMD (38%). Reattack occurred in 2 patients (9.5%) among 21 patients who received surgical revascularization during mean follow-up of 15.1±21.7 months. One patient had ischemic reattack and the other had hemorrhagic reattack after operation. Thirty five (71%) of 50 patients had good outcomes but 3 patients with hemorrhagic reattacks had poor outcomes (6%). **Conclusions** : Probable and definite MMD have some differences in clinical features such as onset age and reattack. Regardless of initial manifestations, most of the adult MMD patients had good outcomes. However, patients with recurrent attacks had worse outcomes than those without reattacks, especially of the hemorrhagic type. Therefore, prevention of rebleeding should receive more attention in the management of adult MMD.

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Key Words : Adult-onset moyamoya disease, Initial manifestation, Reattack, Clinical outcome

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1-3 6~9%가 가 4,5
10 가 30 2 가
80% 10% 1,6,7
60% 5,8
가 가 가 가
가 가

Chi-square, t-

가
 9
 가
 1
 1985 1998
 16 50
 가 가
 10
 definite(bilateral) probable(unilateral)
 (Table 1).

3)

Modified Rankin Scale(MRS: Grade 0, no symptom at all; 1, no significant disability despite symptoms: able to carry out all usual duties and activities; 2, slight disability: unable to carry out all previous activities but able to look after own affairs without assistance; 3, moderate disability: requiring some help, but able to walk without assistance; 4, moderate disability: unable to walk without assistance, and unable to attend to own bodily needs without assistance; 5, severe disability: bedridden, incontinent, and requiring constant nursing care and attention)¹¹ Grade 0-1, 2-3, 4-5 (good), (moderate), (poor outcome) MRS

2. ()
 1)
 가
 가
 (transient ischemic attack) (cerebral infarction) (in-tracerebral hemorrhage) (primary intraventricular hemorrhage)
 2 (probable) (definite)
 2)
 가
 2. (Table 2)
 29 (58%) (20, 40%)
 가
 (Fig. 1). 9 8

1.
 20, 30 1:1.5 가
 17 55
 가
 33±8.7 30 가 (Fig. 1).
 1). (Table 1 criteria 3) 10, 40
 가 2, 8 가 4
 39±10.7 31.5±
 (posterior circulation) 9 (22.5%)
 33 7
 24.4±25.9 10
 3 3, 5, 8
 가 (4%).
 2. (Table 2)
 29 (58%) (20, 40%)
 가
 (Fig. 1). 9 8

Table 1. Angiographic findings for the diagnosis of moyamoya disease (Yonekawa et al., 1992)

1. Stenosis or occlusion is observed at the terminal portion of the intracranial ICA and/or at the proximal portion of the ACA and/or the MCA.
2. Abnormal vascular moyamoya networks are observed in the vicinity of the above-mentioned areas in the arterial phase.
3. These findings are recognized bilaterally.

Abbreviations: ICA, internal carotid artery; ACA, anterior cerebral artery; MCA, middle cerebral artery.

Notes: Definite case, fulfills all the above findings; Probable case, dose not fulfill the criteria 3.

(1) 13, 16 (22%), 1 (36%), 10 (36%), 6 (60%), 11 (18%), 14 (50%), 4.2±4.6, 4, 15 (38%), 15, 20, 39, 12, 3, 2, 21, 20, encephalo-duro-arterio-synangiosis(EDAS), 1, (STA-MCA anastomosis), 20, EDAS, 12 (9, 3), 8 (5, 3), 8.4±4.2, 21, 10, 15.1±21.7 (1, 8, 1, 6), 2 (9.5%), 2, EDAS, 13, 1 가

Table 2. Initial manifestations of adult-onset moyamoya disease

Features	Number	%
Ischemic	29	58
TIA	13	26
Infarction	16	32
Hemorrhage	20	40
ICH	7	14
ICH with IVH	4	8
IVH	9	18
Incidental (facial spasm)	1	2
Total	50	100

Abbreviations: TIA, transient ischemic attack; ICH, intracerebral hemorrhage; IVH, intraventricular hemorrhage.

Table 3. Type of reattack in adult-onset moyamoya disease

First Attack	Second Attack		Reattack rate (%)
	type	number of patients	
Ischemia (N=29)	ischemia	13*	16/29 (55)
	hemorrhage	3*	
Hemorrhage (N=20)	hemorrhage	4*	4/20 (20)
	ischemia	0	
Total			20/49 (41)

* A patient who had episodes more than two was included.

Table 4. Comparison between patients with definite and probable moyamoya disease in adult

	Definite	Probable	p value
Number of Patients	40	10	
Male:Female	18:22	2:8	0.133
Onset Age (years)	31.5±7.7 (17-49)	39±10.7 (17-55)	0.014
Initial Manifestation			
ischemia:hemorrhage	25:14 (1:incidental)	4:6	0.167
Reattack			
ischemia	12	1	
hemorrhage	3	4	
total	15/39 (38)	5/10 (50)	0.058
Recurrence interval (mean±SD, months)	50±63.3	49.8±17.2	

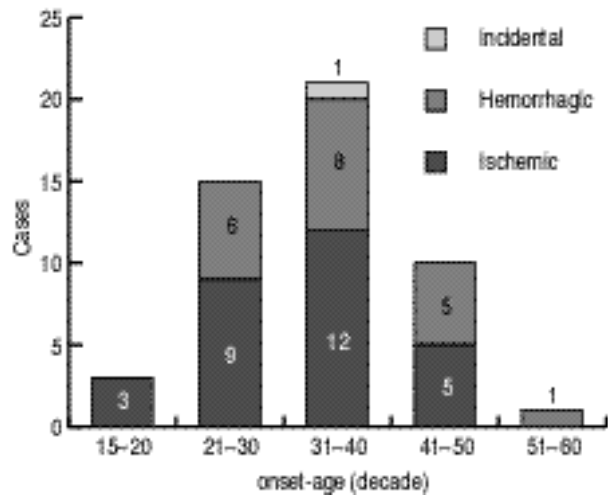


Figure 1. Onset-Age distribution of adult moyamoya disease

STA-MCA anastomosis 13,15-17 가 1

23 2 3 18

, 4 8 가

(Table 5).

4. (Table 6) 22.5% 18

71%가 (89%)

2 81% 59%

가 (striate arteries)

(choroidal artery) medullary artery

3 (38%) 가 8 가 12

bFGF

3 bFGF가 가

Willis

50 2 (4%) 2 bFGF가 (angiogenesis) 5

bFGF가 가

2 bFGF 가 13

가 가 가 6,9

(subependyma)

(moyamoya vessels) (microaneurysm)

basic fibroblast growth factor(bFGF) 1,12 1,19

13,14 가 58% (40%)

2~3 (Table 2)

Table 5. Reattack following revascularization surgery in adult-onset moyamoya disease

Initial Manifestation	Surgical methods		Reattack (%)		
	EDAS	STA-MCA anastomosis	Ischemia	Hemorrhage	Total (%)
Ischemia (N=14)	14	0	0	0	0/14 (0)
Hemorrhage (N=7)	6	1	1	1*	2/7 (29)
Total (N=21)					2/21 (9.5)

Abbreviations: EDAS, encephalo-duro-arterio-synangiosis; STA-MCA, superficial temporal artery-middle cerebral artery.

* The patient had total 4 hemorrhagic episodes and died.

Table 6. Modified Rankin Scale (MRS) according to reattack in adult-onset moyamoya disease

Clinical outcome	without reattack			with reattack*				Total (%)	
	I	H	total (%)	I-I	I-H	H-H	H-I		
Good (MRS 0-1)	10	12	22 (81)	8 [†]	2	2	1	13 (59)	35 (71)
Moderate (MRS 2-3)	3	2	5 (19)	5	0	1 [†]	0	6 (27)	11 (23)
Poor (MRS 4-5)	0	0	0 (0)	0	1 ^{††}	2 ^{††}	0	3 (14)	3 (6)
Total	13	14	27	13	3	5	1	22	49

Abbreviations: I, ischemia; H, hemorrhage.

* Recurrence of post-operation was included.

† A patient with episodes more than two is included †† A dead patient is included

:

(selection bias)

가 10% (Fig. 1). 가 5~80% 가 2~3 15~30% 가 10%가 (leading cause) 41%가 (Table (hemodynamic stress) 가 55%, 20%가 (Table (hemodynamic stress) 가 EDAS 가 1~2 가 (natural history)가 가 4.3% 10%) 63% 가 40 가 3 (Table 4). bFGF 가 10 가 36-43 가 19,29,31,44 가 21 2 (9.5%) (Table 5) 41%(49 20 가 70% 가 81% (Table 6). 가 29%(7 2) , (20%, Table 3) 가 59%가 20% 가 STA-MCA anasto- 3 가 가 3 , 3

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