

ON-LINE APPENDIX

Participants

Patient inclusion criteria were the following: 1) clinical diagnosis of idiopathic MMD according to the consensus criteria and guidelines proposed by the Research Committee on Spontaneous Occlusion of the Circle of Willis, and 2) age older than 20 years. The exclusion criteria were quasi-Moyamoya syndrome associated with conditions such as Down syndrome and neurofibromatosis; cortical infarction and/or subcortical lesions of >8 mm in the largest dimension on conventional MR imaging sequences; intracranial hemorrhage; undergoing revascularization surgery before the study; apparent neurologic deficit due to stroke; or any comorbid illnesses that could affect cognition. After exclusions, 23 patients (6 men and 17 women; 21–58 years of age; mean age, 40.9 ± 9.5 years) were enrolled.

The inclusion criteria for the control subjects were age older than 20 years, no clinical evidence of psychiatric or neurologic disorders, no brain lesions on conventional MR imaging sequences, and currently taking no medications that could affect cognitive function. The control subjects were examined with the Japanese version of the Nelson Adult Reading Test. The mean estimated IQ of these individuals was 108.3 ± 6.6 , according to the Japanese version of the Nelson Adult Reading Test.

Neuropsychological Tests

The Wechsler Adult Intelligent Scale-III provides indices of overall intellectual ability (full-scale IQ, verbal IQ, and motor IQ). Four additional indices were used, which represent the major components of intelligence, namely the Verbal Comprehension

Index, Perceptual Organization Index, Working Memory Index, and Processing Speed Index. The Wisconsin Card Sorting Test measures strategic planning ability, organized search, and use of environmental feedback to shift cognitive sets. Three indicators were calculated, consisting of categories achieved and perseverative errors in Milner and the Nelson. The TMT assesses the speed of information processing (part A) and executive function (part B). The times to complete TMT-A and TMT-B were measured, and the difference in scores between TMT-A and TMT-B (B–A) was calculated. The CPT measures sustained and selective attention and impulsivity, via reaction times and the number of omission errors produced. The Stroop test measures sustained attention via the reaction times to state the text color of displayed color names, where the text color is incongruent with the color name (eg, the word “red” displayed in a blue font). The RST is a complex verbal test that evaluates both storage and processing (ie, reading) elements of working memory. The scores are expressed as the total number and the proportion of words correctly recalled.

Patients were subgrouped into 2, according to their performance scores, error numbers, and reaction time on WCST, CPT, Stroop test, and RST (On-line Table). For the Wisconsin Card Sorting Test and Stroop test, patients with an abnormality in at least 1 subscore were in the impaired test performance group. In TMT, the score of the patient who required >200 seconds for accomplishing tasks was considered not optimal for the quantitative performance evaluation. Accordingly, 1 patient was excluded from the correlation analysis by using Pearson moment-product correlation coefficient.

On-line Table: Results of neuropsychological examinations in patients with MMD

Neuropsychological Examinations	Range (Mean)
WAIS -III	
Verbal IQ	64–121 (95 ± 13)
Motor IQ	71–113 (93 ± 11)
Full-scale IQ	65–120 (94 ± 13)
Subscore	
Verbal perception	61–111 (96 ± 13)
Perceptual organization	55–114 (95 ± 13)
Working memory	58–123 (93 ± 14)
Processing speed	63–133 (96 ± 15)
TMT-A	35–113 (72.2 ± 20.1)
TMT-B	58–217 (93.5 ± 34.8)
B–A	–25–134 (21.2 ± 33.2)
WCST	
CA (<4) ^a	1–6 (4.9 ± 1.4)
PEM (>2) ^a	0–6 (1.7 ± 1.8)
PEN (>3) ^a	0–15 (2.6 ± 3.7)
Stroop test (>10) ^a	2–129 (14.3 ± 26.7)
CPT	
Error (>3) ^a	0–71 (6 ± 14.8)
RT (>480 ms) ^a	312.7–1533 (494 ± 250.6)
RST	
(<2) ^a	1.5–4 (2.5 ± 0.6)

Note:—WAIS-II indicates Wechsler Adult Intelligent Scale-III; CA, categories achieved; B–A, difference in scores between TMT-A and TMT-B; PEM and PEN, perseverative errors in Milner (PEM) and Nelson (PEN) tests; RT, reaction time (sec); WCST, Wisconsin Card Sorting Test.

^a Performance scores, error numbers, or reaction time, which are considered cutoff values of impaired neuropsychological performance.