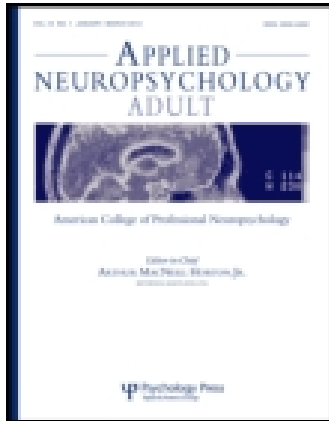


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Moyamoya Disease: A Review of the Literature

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Moyamoya Disease: A Review of the Literature

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Moyamoya disease is a rare progressive cerebrovascular disorder that can be congenital or acquired and involves progressive stenosis and occlusion of cerebral arteries. The brain through compensatory angiogenesis then attempts to compensate for ischemia by producing a local network of tiny blood vessels, which appear cloud-like on angiograms. Consistent with multifocal or diffuse areas of ischemia, people with moyamoya often have multiple areas of cognitive impairment. A thorough literature review of the neuropsychological profile in individuals diagnosed with moyamoya disease is provided. Medical and neuropsychological/neurobehavioral data of a middle-aged woman with moyamoya disease is also described. The patient (MA) suffered an embolic shower with insult to both cerebral hemispheres. Neuropsychological results suggested a multifocal decline, with both cortical and subcortical involvement. Data were consistent with impairments in attention, concentration, executive skills, processing speed, and memory encoding and retrieval, with relatively spared aspects of memory and language skills. This case study supports the literature and provides an additional example of the neuropsychological profile and deficit pattern of an individual with moyamoya following an embolic stroke shower.

Key words: cerebrovascular disorders, moyamoya disease

Moyamoya disease is a rare congenital or acquired cerebrovascular disorder of unknown etiology (Jefferson, Glosser, Detre, Sinson, & Liebeskind, 2006; Moore, Lee, & Macciocchi, 1997; Setzen et al., 1999; Yamada, Nakagawa, Matsushima, & Shibuya, 2001). Several diseases are possibly linked to moyamoya and associated abnormal vasculature, including hematological disorders (e.g., sickle cell anemia, lupus) and immunological conditions (e.g., Graves disease/thyrototoxicosis).

Infections (e.g., leptospirosis, tuberculosis), congenital disorders (e.g., Down syndrome, Marfan syndrome, Turner syndrome), and vascular disease (e.g., atherosclerotic disease) have also been implicated as potential etiologies of moyamoya-type vascular abnormalities (Hallemeier et al., 2006). Moyamoya disease is characterized by progressive stenosis and/or occlusion of the distal internal carotid arteries and the circle of Willis. The disease typically causes stenosis and occlusion of small blood vessels and resulting ischemia and hypoxia. The ischemia drives massive angiogenesis that produces numerous but tiny branched collateral blood vessels. This massive compensatory angiogenesis results in a

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thick mesh of tiny blood vessels, which appear cloud-like or “smoke-like” on cerebral angiography. These are often referred to as “moyamoya vessels,” which loosely means “puff of smoke” in Japanese. Stenosis or occlusion can often be seen in portions of the internal carotid artery extending toward the more proximal portions of the middle (MC) and anterior cerebral (AC) arteries (Yamada, Himeno, Suzuki, & Matsushima, 1995). Not only are the MC and AC arteries implicated, but the posterior cerebral artery is involved around 30% of the time (Kuroda, Ishikawa, Houkin, & Iwasaki, 2002). Moyamoya is typically first seen on a computed axial tomography (CT) scan, magnetic resonance imaging (MRI), or angiography following migraine-like headaches, cerebrovascular events, or convulsions.

Moyamoya disease was first described in 1957 and was thought to be primarily a disorder affecting Japanese children (Suzuki & Takaku, 1969). However, more recent research has shown increasing numbers of adult patients of all nationalities. Epidemiologically, it is estimated that 10.5 patients per 100,000 people in Japan have moyamoya disease, with a 2.18 ratio of female-to-male patients (Baba, Houkin, & Kuroda, 2008). Although there are relatively few studies examining prevalence in the United States, one study examining prevalence in California and Washington found 0.086 patients per 100,000 with a ratio of 4.6 Whites to Asian Americans, 2.2 Whites to African Americans, and 0.5 Whites to Hispanics (Uchino, Johnson, Claiborne, & Tirschwell, 2005). The age distribution appears bimodal—that is, either children younger than 10 years of age or adults aged between the third and fifth decades are affected (Chiu, Shedden, Bratina, & Grotta, 1998). Hallemeier et al. (2006), in their review of 34 adults diagnosed with moyamoya, determined a median age of 42 years; 73.5% were women. There was also a high incidence of recurrent stroke, especially with bilateral vascular involvement.

Moyamoya is hypothesized to be familial, especially in Japan (Takagi, Kikuta, Nozaki, & Hashimoto, 2007). Research has suggested that there is a 10% higher prevalence rate in Japanese and Koreans, thus implying genetic inheritance (Kitahara, Ariga, Yamaura, Makino, & Maki, 1979). Additionally, current genetic studies implicate chromosomes 3, 6, and 17 (q25.3); however, further research is needed to clarify genetic and other risk factors (Fukui, Kono, Sueishi, & Ikezaki, 2000; Mineharu et al., 2008).

When the carotid arteries become significantly stenotic or there are recurrent transient ischemic attacks, individuals with moyamoya experience reactive vascular changes, which may include a cerebrovascular accident or an embolic shower (Takagi et al., 2007). This stenosis rarely progresses in adults but often advances in children; thus, adults may go without symptoms for

long periods (Shirane, Mikawa, & Ebina, 1999). When symptoms do appear, children are likely to present with cerebral ischemia or infarction, and adults typically show signs of intracerebral hemorrhage (Chaudhuri et al., 1993). Some research suggests significant diversity in clinical presentations, which may range from nearly asymptomatic to significantly impaired (Jefferson et al., 2006).

Given high diversity in symptoms as is related to variability in etiology (stroke, embolic shower, etc.), a comprehensive neuropsychological assessment is recommended rather than focal evaluation. This approach will best allow the provider to examine strengths and weaknesses so as to assist in treatment planning, better predict outcome, and guide rehabilitation, regardless of whether or not assessment is done presurgery or post-surgery. Additionally, assessment should include evaluation of mood, given potential difficulties with adjustment to medical illness or the direct effects of cerebral compromise on psychiatric functioning. Similar to the overall neurocognitive profile, symptoms of mood tend to vary depending on location of injury and situation (presence of support system, etc.).

Neuropsychological Preoperative Sequelae

The neuropsychological sequelae associated with moyamoya appear to be reliant on the areas of the brain involved, severity of the vascular event, age of onset, and the type and success of treatment (Setzen et al., 1999). Up to two thirds of patients appear to have neurocognitive impairment (Festa et al., 2010). Case reports during recent decades have emerged, including both pediatric and adult populations. Though adult studies typically do not find significantly diminished Full-Scale IQ (FSIQ), research on children with moyamoya reported significantly reduced and often below-average FSIQ in addition to the multifocal presentation (Williams et al., 2012). Individuals diagnosed with moyamoya who have undergone neuropsychological evaluations prior to surgical intervention have demonstrated various cognitive impairments across the domains of intelligence, processing speed, executive dysfunction, and visual-spatial abilities (Bornstein, 1985; Hoare & Keogh, 1974; Jefferson et al., 2006; Lubman, Pantelis, Desmond, Proffitt, & Velakoulis, 2003; Ogasawara et al., 2005). For example, Ogasawara et al. (2005) investigated the neurocognitive functioning of a 48-year-old man. Preoperative testing revealed impaired Performance IQ (PIQ = 68), average Verbal IQ (VIQ = 97), an FSIQ the low-average range (FSIQ = 83). Jefferson et al. (2006) documented the case of a 48-year-old woman revealing strong language and verbal reasoning abilities; however, this woman had suffered a right-hemisphere stroke 7 months prior to testing

and this impacted her visuospatial perception, organization, and construction. Particular impairment also was noted during tasks assessing spatial relationships, nonverbal visuospatial memory, and verbal learning measures. One empirical study presented neuropsychological data on 26 adults with moyamoya disease prior to any surgical intervention (Festa et al., 2010). Most (66%) exhibited mild-to-moderate cognitive impairments, which were defined in this study as more than one standard deviation below mean scores. Most individuals had multifocal cognitive dysfunction, and the relative frequencies of cognitive skills affected were as listed: manual dexterity (58%), grip strength (36%), verbal memory (31%), processing speed (29%), verbal fluency (26%), and executive functioning (25%). One of the largest moyamoya studies involving 36 adults with no previous history of neurosurgical intervention revealed an incidence of cognitive impairment at 31% ($n = 11$ of 36; Karzmark et al., 2008). The majority of deficits fell into the mild impairment range. In sum, it appears that preoperatively, individuals with moyamoya exhibit broad cognitive impairments.

Surgical Intervention

Prognostic factors include bilateral vasculopathy, associated strokes, and duration of disease process (Williams et al., 2012). The surgical treatment is common, and a variety of procedures are utilized. Encephaloduroarteriosynangiosis, encephalomyosynangiosis, external carotid–internal carotid bypass surgery, superficial temporal artery–MC artery bypass surgery, and burr hole placement are potential surgical interventions; all of these surgeries are designed to either remove, indirectly revascularize, or circumvent abnormal vasculature (Fung, Thompson, & Ganesan, 2005; Kinugasa, Mandai, Kamata, Sugi, & Ohmoto, 1993; Takagi et al., 2007). Prognosis varies greatly; however, with successful vascular surgery, prognosis can be good, especially if an early diagnosis is made (Hallemeier et al., 2006).

Neuropsychological Postoperative Sequelae

The literature suggests that investigations of moyamoya cases following surgical interventions have been variable. The majority of reports demonstrate improvement in cognitive deficits after surgical intervention. For example, neuropsychological testing postoperatively in the 48-year-old case presented by Jefferson et al. (2006) revealed improvement of visuospatial, nonverbal visuospatial memory, and verbal learning and delayed recall performances. However, unfortunately, not all cases retain premorbid or average cognition following surgery. Bowen, Marks, and Steinberg (1998) reported two pediatric case studies (5-year-old boy and 7-year-old girl) in

which both children continued to show gross cognitive deficits with significant limitations evidenced in blind tactile-spatial skills and motor function following bilateral revascularization. Follow-up of the 48-year-old man reported by Ogasawara et al. (2005) revealed that at his 3- and 6-month evaluations following artery bypass, the majority of neurotesting remained consistent (FSIQ = 86/88). Digit Span testing went from average (scaled score [ss] = 8) to impaired ($ss = 3$) indicating significant decompensation in working memory and attention. Executive dysfunction appears to be a domain of specific concern within the moyamoya population and was found in 60% ($n = 6$ out of 10) of cases when investigated by Calviere et al. (2010). A correlation was made between executive dysfunction and reduced regional cerebral vascular reserve in the frontal areas. Calviere et al. also found verbal memory to be impaired in 50% of their patients postoperatively. Frontal-lobe dysfunction was also presented in a single case study by Schwarz, Thurstin, and Levine (2011) who investigated a 25-year-old woman following revascularization surgery and ischemic stroke at age 15. Assessment data revealed specific deficits in inhibition and problem-solving. Although it appears overall that cognitive functioning increases following surgical intervention in moyamoya cases, the neuropsychological postoperative profile remains variable.

CASE BACKGROUND

This case study presents the neuropsychological data from an evaluation of a 42-year-old right-handed Caucasian woman (MA) following a brain-imaging-confirmed embolic stroke shower and diagnosis of moyamoya. While in the hospital, the patient completed multiple neuropsychological measures, including the *Beck Depression Inventory-II* (BDI-II; Beck, Steer, & Brown, 1996), *Boston Naming Test* (BNT; Kaplan, Goodglass, & Weintraub, 1983), *California Verbal Learning Test-Second Edition* (CVLT-II; Delis, Kramer, Kaplan, & Ober, 2000), *Controlled Oral Word Association Test* (COWAT, Neurosensory Centre Comprehensive Examination for Aphasia; Spreen & Benton, 1977), *Multilingual Aphasia Examination-Seventh Edition* (MAE-VII; Benton, Hamsher, & Sivan, 2000), *Rey-Osterrieth Complex Figure Test* (Rey & Osterrieth, 1944), *Repeatable Battery for the Assessment of Neuropsychological Status* (RBANS; Randolph, 1998), *Stroop Color and Word Test* (Stroop; Golden & Freshwater, 2002); *Trail-Making Test* from the *Halstead-Reitan Neuropsychological Test Battery* (TMT-A and TMT-B; Reitan & Wolfson, 1993), *Visual Organization Test* (VOT; Hooper, 1983), *Wechsler Abbreviated Scale of*

Intelligence (WASI; Wechsler, 1999a), *Wechsler Memory Scale-Third Edition* (WMS-III; Wechsler, 1999b), and *Wechsler Test of Adult Reading* (WTAR; Wechsler, 2001).

MA was admitted to the hospital following an acute onset of left-sided weakness and imbalance. MA reported experiencing a similar event approximately 3 months prior, characterized by numbness of the left face, arm, and leg. Additionally, MA told of a month-long progressive cognitive decline characterized by “spacing out,” poor attention and concentration, impulsivity, increased irritability, difficulty encoding new information, and word-finding difficulties. The symptoms were subsequently found to be secondary to a series of strokes due to what was ultimately determined to be moyamoya disease. While in the hospital, several behaviors were observed that corresponded with her report: difficulty sustaining attentional focus, frequent loss of her train of thought, verbal impulsivity, an inability to inhibit her responses on a go–no go task, and distractibility. When distracted, MA was unable to inhibit an opposite hand response. Further, she was also unable to keep her eyes closed when instructed to do so and stated, “I’m trying, they just want to open.”

Prior to her embolic stroke shower, MA was living with her fiancé, working at a fast food restaurant, and independently managing basic and instrumental activities of daily living. MA completed 12 years of education with no reported history of special education, developmental disorders, or learning difficulties. Medical history was significant for headaches, leukocytosis, iron-deficiency anemia, hypertension, and moyamoya disease (diagnosed following neuroimaging conducted at the time of this evaluation). MA denied use/abuse of nicotine, alcohol, or recreational drugs. MA denied any past motor, sensory, or cognitive declines, other than those previously described. Psychiatric history was negative. Family history was significant for moyamoya disease in her sister. Perhaps the combination of genetics and hypertension predisposed this patient to moyamoya disease. At the time of the assessment, medications included acetaminophen, aspirin, bisacodyl, enoxaparin, fexofenadine, ondansetron, promethazine, Senokot, and simvastatin. MA was evaluated when she was cognitively able to effectively participate in the evaluation, approximately 1 week after admission.

Neuroimaging was consistent with a diagnosis of moyamoya disease. See Figure 1. Cranial CT perfusion demonstrated cerebral ischemia to both the frontal and parietal lobes in the anterior circulation supplied by the MC arteries, with greater evidence noted in the right hemisphere. Arteriogram showed extensive vascular abnormalities including, but not limited to, severe near occlusive narrowing of the right internal carotid artery, reduced right MC artery circulation, a small right

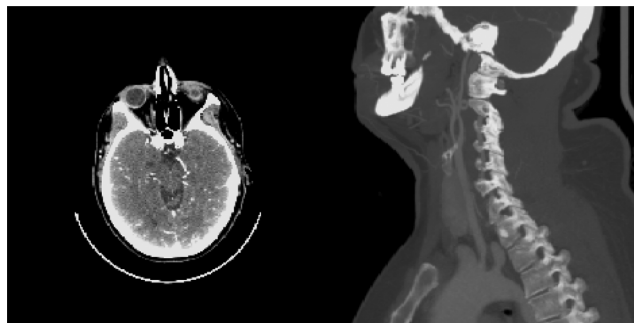


FIGURE 1 Neuroimaging (CT of the head and angiogram of the neck) of the patient demonstrating the effects of the embolic shower and vascular changes to the carotid arteries consistent with moyamoya disease.

posterior communicating artery, and a typical moyamoya mesh of tiny abnormal lenticulostriate collaterals.

CT angiogram of the head and neck revealed an acute bilateral corpus callosum infarct, severe narrowing of both supraclinoid internal carotid artery segments as well as both M1 MC artery segments and the right A1 AC artery segments. Additionally, the internal carotid arteries were abnormally small, mild luminal narrowing was noted on the proximal aspect of the left internal carotid suggesting chronic dissection, and numerous entangled lenticulostriate vessels were present bilaterally.

MRI of the head noted an acute infarction involving the corpus callosum, cortical grey matter in the right cerebral hemisphere, and subcortical white matter of the right hemisphere consistent with an embolic shower. Ultimately, moyamoya disease was diagnosed by the neuroradiologist, and the mesh or netlike nature of the collaterals was interpreted as being very consistent with moyamoya.

TEST RESULTS AND SUMMARY

Based on her education level, stable employment history, and multiple scores in the average range, it is suspected that MA is likely of average premorbid general intellectual ability (see Table 1). Her WTAR and vocabulary are felt to be underestimates of her premorbid ability level. MA appears to have grossly intact and average scores on measures involving auditory working memory, aspects of verbal memory functioning (Story Memory, Logical Memory II, Visual Reproduction Repetition), and some measures of object naming. MA had an average performance on the 10-item object-naming subtest of the RBANS but had a borderline range score on the more demanding BNT. Data also revealed relatively spared areas of language, such as receptive language, thus allowing her to function, at least verbally, close to her premorbid borderline level of intellectual functioning; this is likely secondary to a more intact left hemisphere, as noted on neuroimaging.

TABLE 1
Neuropsychological Assessment Results

<i>Test</i>	<i>Score</i>	<i>Description</i>
<i>WTAR</i>		
Total Score	Std score = 77	Borderline
<i>WASI</i>		
Vocabulary	Std score = 60	Profound
<i>BNT</i>		
BNT	Std score = 78	Borderline
<i>MAE-VII</i>		
Token Test	Std score = 80	Low Average
<i>RBANS</i>		
Total Scale	Std score = 59	Profound
Attention	Std score = 72	Borderline
Digit Span	Std score = 96	Average
Coding	Std score = 52	Profound
Immediate Memory	Std score = 81	Low Average
List Learning	Std score = 57	Profound
Story Memory	Std score = 101	Average
Delayed Memory	Std score = 48	Profound
List Recall	Std score = 66	Mild
List Recognition	Std score < 45	Profound
Story Recall	Std score = 76	Borderline
Figure Recall	Std score = 52	Profound
Language	Std score = 76	Borderline
Picture Naming	Std score = 108	Average
Semantic Fluency	Std score = 65	Moderate
Visual-Spatial/Visual-Construction	Std score = 60	Profound
Figure Copy	Std score < 45	Profound
Line Orientation	Std score = 73	Borderline
<i>COWAT</i>		
F	Total raw: 3	
A	Total raw: 2	
S	Total raw: 2	
Total	Std score < 1	Profound
Animals	Std score < 45	Profound
<i>Stroop Color and Word Test</i>		
Color Reading	Std score < 55	Profound
Word Reading	Std score < 55	Profound
Color-Word	Std score < 55	Profound
<i>Trail-Making Test</i>		
A	60	
B	Unscorable—too impaired	
<i>CVLT-II</i>		
Short-Delay Free Recall	Std score = 55	Profound
Short-Delay Cued Recall	Std score < 45	Profound
Long-Delay Free Recall	Std score < 45	Profound
Long-Delay Cued Recall	Std score < 45	Profound
Total Rec. Discriminability	Std score < 45	Profound
<i>WMS-III</i>		
Logical Memory I	Std score = 74	Borderline
Logical Memory II	Std score = 91	Average
Visual Reproduction I	Std score = 60	Profound
Visual Reproduction II	Std score = 74	Borderline
Visual Reproduction Recognition	Std score = 90	Average
<i>Rey Complex Figure Test</i>		
Immediate Trial	Std score < 55	Profound
Delayed Trial	Std score < 55	Profound
Recognition	Std score = 66	Mild
<i>VOT</i>		
Total	Std score = 73	Borderline
<i>Word Memory Test</i>	Indicative of good effort engagement	
<i>Test of Memory Malingering</i>	Indicative of good effort engagement	
<i>BDI-II</i>	Raw score = 15	Mild

Neuropsychological test results suggested mild impairments on some measures of information encoding, attention skills, recall of visual information, and visual organization. Her memory impairments were more severe with less organized and nonsemantic information. Examination of attention and executive function provided evidence for both cortical and subcortical involvement. Attention skills performance greatly decreased as tasks became more complex, and a measure involving alternating attention (TMT-B) had to be discontinued due to inability to shift attention effectively. Similarly, information-processing speed appeared impaired as well on the measures administered.

Executive functions were impaired, both on testing and during the neurobehavioral exam. The Stroop, COWAT, TMT-A, and TMT-B were all severely impaired, though imaging did not show significant prefrontal involvement. In fact, the imaging really contrasts with both the clinical presentation and impaired executive function testing. This contrast may highlight the distributed nature of executive function systems. MA was easily distracted, frequently lost her train of thought, and was unable to inhibit her responses. Testing suggests severe deficits in mental organization and inhibition. These findings are consistent with previous research showing executive dysfunction in moyamoya (Karzmark et al., 2008; Lubman et al., 2003). She was better able to consolidate and recall preorganized verbal information and simple rather than complex geometric figures. However, consolidation and recall of a word list and a complex geometric figure were both severely impaired, likely as a consequence of more anterior compromise and difficulty organizing information so as to aid in recall. Visuospatial abilities revealed mildly impaired visual organization and perceptual abilities, which provide further evidence for focal areas of deficit. These findings may be related to the right-hemisphere cortical and subcortical pathology and narrowed arteries as reported on imaging. Additionally, brief assessment of mood revealed mild depression (BDI-II). Following this assessment, MA was transferred to a setting where she reportedly underwent cerebrovascular surgery for revascularization. Her outcome is not known, but outcomes can vary dramatically, both postsurgically and untreated.

CONCLUSIONS/IMPLICATIONS

This purpose of this article was twofold. First, we set out to review the literature on moyamoya disease to present an accumulation of current findings. Secondly, we presented neuropsychological information of an adult case as an additional example of the course and manifestation of moyamoya disease. As shown in the literature on moyamoya disease along with the current case presented, the utility of neuropsychological assessment

is beneficial in clarifying specific areas of deficit. This is apparent from the consistency of neuropsychological profiles between cases, particularly preoperatively. The literature review revealed broad cognitive impairments including intelligence, processing speed, visual-spatial performance, and executive functioning (Bornstein, 1985; Festa et al., 2010; Hoare & Keogh, 1974; Jefferson et al., 2006; Karzmark et al., 2008; Lubman et al., 2003; Ogasawara et al., 2005). Coherent findings were shown during our case evaluation where MA demonstrated impairments on measures of information encoding, attention skills, recalling visual information, visual organization, processing speed, and many aspects of executive functioning. Mild depression was also noted. The neuropsychological findings from this case also align with imaging reports that have been presented in many of the previous cases across the literature. Consistent with the multifocal vessel occlusion and the brain's attempt to compensate, the current case's neuropsychological test results provided evidence of multifocal decline in both cerebral hemispheres, with what appears to be both cortical and subcortical involvement, consistent with a multi-infarct etiology. Consistent with previous research, there was some sparing of memory and language functions. Overall, this literature review provides the field with an overview of the neurocognitive findings and neuropsychological profile to date in those diagnosed with moyamoya disease. The presented case demonstrates the quick course of progressive multifocal symptoms that can be differentiated from other vascular events. Given the treatable nature of this condition, such symptom expression should continue to prompt neuropsychologists to make appropriate referrals for neurological consultation and imaging to clarify the etiology. A limitation of this case was the lack of postoperative neuropsychological data available. A reevaluation of the case postoperatively would have been beneficial.

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