Serial Neuropsychological Assessment of an Adult with Moyamoya Disease and Borderline Personality Disorder

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INTRODUCTION

Moyamoya disease (MMD) is a rare cerebrovascular condition characterized by progressive stenosis of the internal carotid arteries (ICA) and their proximal branches. The abnormal moyamoya vessels take on the appearance of a “puff of smoke” on angiogram, which is connoted by the Japanese term moyamoya.

Whereas MMD is most common in individuals of Asian descent, it has been observed in other ethnicities. In North America, its prevalence has been estimated at 0.086 per 100,000 persons. Females are generally twice as likely to have MMD as males. Peak onset/incidence is bimodal and highest in children (~5-10 years) and adults (~30-40 years). Though its etiology is unknown, both genetic and stress-diathesis models have been proposed.

Individuals with MMD are predisposed to hypoperfusion, transient ischemic attack, seizure, ischemic stroke, and cerebral hemorrhage. Stress, fatigue, infection, dehydration, and substance use may precipitate ischemic symptoms. Hemorrhage may result from moyamoya vessels dilating and rupturing from hemodynamic stress, and the development and rupture of saccular aneurysms. Re-vascularization surgery (e.g., redirecting blood flow via the spared external carotid) is the preferred mode of intervention; it is associated with reduced mortality, good cognitive outcomes, and a 96% probability of remaining stroke-free 5 years post-surgery.

Of the neuropsychological phenotypes described in the literature, cognitive functioning pre- and post-stroke is fairly well documented. In pre-stroke MMD, frontal-subcortical deficits are common. Post-stroke cognitive outcomes tend to vary with stroke location, type, severity, and mode of treatment (conservative treatments fare worse than surgery).

In contrast, the prevalence and types of neuropsychiatric sequelae in MMD are less clear. A few studies have described post-stroke depression/anxiety. According to several published case reports, new onset psychosis and manic symptoms in otherwise healthy individuals have prompted neuroimaging that subsequently led to MMD diagnoses. Longstanding personality disorder, on the other hand, has not been reported in the MMD literature.

The following case describes a young woman with newly diagnosed MMD, status-post stroke, presenting with acute neuropsychiatric symptoms overlaid on longstanding borderline personality traits.
CASE HISTORY

The patient was a 27-year-old, right-handed, adopted woman of mixed race. Her upbringing was tumultuous. Prior to her adoption during grade school, she had lived with several foster families and suffered sexual trauma. She reportedly ran away from home on multiple occasions and she dropped out of school in the 8th grade. Her past psychiatric history was notable for labile moods with feelings of “self-hatred” since 12 years of age. While she initially denied suicidal ideation and attempts, a collateral informant said that she occasionally manipulated significant others with nonspecific threats of self-harm. Her past medical history was remarkable for herpes simplex virus (type 2) and hypertension. Prominent psychosocial issues in her adult life included prostitution, polysubstance abuse with opioid dependence, and recent child protective services involvement with her toddler.

The patient initially presented to the emergency department (ED) complaining of what she believed to be opioid withdrawal symptoms over the previous few days. She was sent home with Suboxone and outpatient referrals. However, she returned to the ED the next day with left-sided weakness and blood pressure of 170/105. A CT confirmed acute-on-chronic occlusion of her right middle cerebral artery (MCA). Subsequent MRIs revealed restricted diffusion and ischemia of right frontal, temporal, and parietal lobes (Figure 1). RAPID MRI appreciated a perfusion diffusion mismatch suggestive of MMD. Angiogram ultimately confirmed MMD, with involvement of the right ICA extending prominently into the right M1 segment (Figure 2).

She was levetiracetam loaded for seizure prophylaxis and admitted to neurocritical care. Her admission was marked by frequent agitation and treatment noncompliance; she perseverated on returning home and leaving the hospital against medical advice. Psychiatry consulted on the case and discontinued levetiracetam in case her behavior was due to “Keppra Rage.” She refused mood stabilizers. Four-point restraints were threatened and a 1:1 security guard was posted in her room – all to no avail. Subsequently, inpatient neuropsychological consultation was ordered to aid in clinical decision-making and discharge planning.
Upon our initial approach, the patient was standing in her hospital room yelling at security and nursing staff. She was tearful, profane, and demanding that she be discharged home to care for her child. After validating her concerns and explaining the rationale that right hemisphere stroke often results in “anosognosia” or compromised awareness of one’s stroke-related difficulties, she grew more amenable to limit setting and appeals that her behavior was counterproductive to her goals. She ultimately confided that she did not appreciate the changes others were noticing in her. Her adoptive sister was interviewed and stated that although the patient was pre-morbidly labile, sexually provocative, impulsive, and manipulative, her current level of agitation/hostility was more pronounced and her insight and judgment were diminished. The patient eventually agreed to cognitive testing, which commenced two hours later so that she would have sufficient time to calm down.

When the examiner returned, he stood in the patient’s left field of vision but she did not notice him. She had left-sided facial droop, right-sided gaze preference, and left-sided foot drag. Her speech was pressured and dysarthric. Her thought process was tangential. Thought content centered on securing a home discharge. Interpersonal boundaries were loose, as she made frequent ingratiating statements to the examiner and compared him to other providers whom she devalued. Her stated mood varied from “great” to “worthless.” She interrupted frequently and was often distracted mid-task. She had limited awareness of these tendencies.

On testing, she did not know the correct day, month, or date. She exhibited severely impaired attention and marked visuospatial/constructional deficits with left-sided neglect (Figure 3). These impairments undermined performance on executive and visual memory measures. Language and verbal learning/memory storage were grossly intact but with variable delayed verbal recall. Insight and judgment/decision-making were poor. Following feedback about her cognitive test results and their implications for her health, independence, and ability to safely parent her child, she agreed to start mood stabilizers, gave her sister Power of Attorney, and accepted a discharge to a skilled nursing facility.
Six months later, however, the patient was re-admitted with new left MCA infarcts in the caudate head and subcortical white matter. Since her prior hospitalization, she was evicted from her apartment, arrested on an outstanding warrant, lost custody of her child, and attempted suicide. Neuropsychological re-evaluation was ordered to establish a new baseline prior to her undergoing left-sided re-vascularization surgery.

On re-assessment, though she demonstrated some increased awareness of her cognitive/behavioral impairments, her overall functioning remained negatively impacted. She reported ongoing anger management problems (worse than before her strokes) and difficulties with everyday judgment/problem solving (e.g., moving into a garage with no heat after her eviction in mid-winter rather than asking her sister for help). She mentioned an instance of nearly causing a fire after she forgot food cooking in the oven. She also noted that she frequently called her sister at inappropriate times of night despite being discouraged from doing so. Ongoing issues with visual neglect were also endorsed (e.g., getting out of a car without realizing her seatbelt was still fastened).

Her left-sided facial droop was less pronounced and her gait had normalized. Her left hand was somewhat contracted and weaker than her right. Visual field testing revealed variable attention to the left hemispace and extinction to the left side with simultaneous visual stimulation. Her speech was aprosodic but with clear articulation and normal rate and volume. Her thought process was circumstantial. She continued to display labile affect and loose boundaries; however, these were also somewhat improved. Her mood varied from “depressed” to “hopeful.” She denied suicidal ideation, intent, and plans.

On testing, she demonstrated improvements in orientation, basic attention, left-sided neglect, and processing speed. She continued to exhibit marked visuospatial impairment and variable memory recall with grossly intact recognition memory. Whereas formal testing of her executive functions was previously precluded by her attentional/visuospatial deficits, she was now testable but showed impaired cognitive flexibility. Although her re-hospitalization was initiated due to new left MCA infarcts, she did not exhibit language impairment.

She felt encouraged by the feedback that many of her initial deficits were recovering and accepted that her residual impairments and limited insight into these deficits were contributing to her day-to-day problems. She was receptive to recommendations for continued supervision/support following discharge and the need for an auxiliary decision maker. She underwent left-sided re-vascularization several days later; however, her post-operative course was complicated by re-emergence of disinhibition, agitation, treatment non-compliance, and suicidal ideation. Once she was medically stable, she was transferred to inpatient psychiatry for further management.

COMMENTARY

On the one hand, this patient’s initial presentation and persisting deficits (i.e., contralateral neglect, disinhibition, and anosognosia) highlight classic neuropsychological sequelae of right MCA infarction. While her low level of formal
education may have contributed to her neuropsychological test performance, it cannot fully account for the extent and quality of her impairments. It is impossible to disentangle whether her borderline personality traits developed independently of MMD, were potentially mediated by chronic hypoperfusion, or represented a neuropsychiatric prodrome of her MMD induced strokes. Regardless, her longstanding behavioral problems were exacerbated by stroke, and led to worse overall functioning and prognosis. It is curious that her more florid manic symptoms manifested in the context of acute right-sided ischemia and re-emerged following re-vascularization. This is especially interesting in light of research proposing a possible shared neurobiological basis of autosomal dominant MMD and bipolar disorder on chromosome 17q25.3 (Mineharu et al., 2008).

This case also highlights the importance of involving neuropsychologists in the management of neurologically/psychiatrically complex patients. A detailed clinical history, serial cognitive assessment, and multidisciplinary communication were critical in informing this patient’s prognosis and addressing her long term needs. Neuropsychology continues to consult with her treating providers and family, and has offered psychoeducation and made specific recommendations about community services, compensatory techniques, and other strategies to help manage her behavior. The patient has since moved in with family, enrolled in substance abuse treatment, and reportedly has maintained sobriety thus far. She is scheduled to undergo right-sided re-vascularization surgery in the near future.

**SELECT REFERENCES AND RECOMMENDED READING**


