Case Report

DOI: 10.6966/EDMJ.202203_9(1).0003

Psychiatry / Neurology Interface: Moyamoya Disease in a Woman with Bipolar Disorder

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Moyamoya disease (MMD) is a rare but often severe cerebrovascular disorder that damages the parenchyma of the brain and might present with uncommon neuropsychiatric symptoms. We report a middle-aged woman with bipolar disorder who manifested manic symptoms as a neuropsychiatric comorbidity of her newly diagnosed MMD. She also presented with vague headache, modest cognitive deficits and a relatively high sensitivity to psychotropic-related adverse effects. Clinical recovery was noted within a month following individualized treatment including psychotropics and conservative treatment for MMD. Although non-specific somatic complaints (especially soft neurological signs) are common in patients with major mental illnesses, the importance and reliability of such complaints are usually underestimated. "Atypical" clinical features of mental illnesses and unusual treatment response should alert clinicians to the possibility of an organic origin to avoid a substantial delay in diagnosis.

Key words: Moyamoya disease, mania, neuropsychiatric symptoms

Introduction

Moyamoya disease (MMD), which is characterized by progressive stenosis of the terminal parts of bilateral internal carotid artery (ICA) and its branches, is a rare cause of stroke and may present with haziness of dilated collateral vessels on angiography. MMD was initially described in Japan with the incidence higher in Japan and China than in Taiwan and North America.[1] Neuropsychiatric morbidities associated with MMD are uncommon, and mostly reported as isolated cases. There was a wide variation in the psychotic symptoms associated with MMD. A previous study has reported a patient presenting with manic symptoms as well as facial palsy and other neurological signs,[2] while irritability, suspiciousness, headache and other cognitive deficits were noted in another patient.[3] Besides, a case of Capgras syndrome (i.e., a delusional belief that a familiar person has been replaced by an imposter) has been described.[4] We present a 45-year-old woman who manifested with manic symptoms as well as atypical clinical features and treatment responses that prompted further investigations and the subsequent diag-

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Received: December 3, 2020 Accepted: January 14, 2021
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nosis of MMD.

Case Report

A 45-year-old married woman, with a psychiatric history of bipolar I disorder, was sent involuntarily to our emergency department due to the chief problem of acute-onset emotional and behavioral disturbance for several days. Tracing her psychiatric history revealed an age of onset around 25 years with initial presentations of abnormal and persistent elevated mood, psychomotor agitation, hyperactivity, sleep disturbance and violent behavior. The presence of associated psychotic symptoms such as delusion or hallucination could not be confirmed. For her personal history, there was a remarkable finding at the age of 15 when she suffered from a sudden-onset weakness over her right-sided extremities for about fifteen minutes before regaining her muscle power. However, she never sought any medical advice or treatment. Besides, she denied illicit drug or alcohol use, related family history, or major surgeries. Although mental status examination on acute psychiatric ward admission showed a clear sensorium, distracted attention and fluctuating euphoric mood with labile affect, occasional restlessness, interrupted and incoherent speech with shift in topics, flight of ideas (mainly vague somatic complaint such as abdominal, neck, and back pain as well as headache), and mild disorientation without paranoia or perceptual distortion were noted. Neurological examination showed a Grade 5 muscle power in all limbs with other systems being normal. Laboratory studies confirmed hyperlipidemia. After intense psychopharmacological and behavioral treatment for three weeks, the patient became more cooperative after stabilization of her mood symptoms. However, she demonstrated a high sensitivity to extrapyramidal symptoms associated with antipsychotic treatment during hospitalization. Parkinsonism with bradykinesia as well as upper limb rigidity and clumsiness developed after the prescription of daily risperidone at a dose of 3 mg for five days. As a result, a “start low, go slow” dosing strategy was started with quetiapine 100 mg and valproate 1,000 mg daily. After treatment, neuropsychological assessment of her cognitive function still revealed very slow response with prolonged processing, poor concentration, impaired working memory and executive functioning as well as poor verbal fluency. Nevertheless, most somatic complaints were improved despite persistent vague headache. Electroencephalographic examination showed intermittent 20 – 50 μV slow waves (4 – 5 Hz) in the right cerebral region with involvement of the temporal and occipital areas, suggestive of a regional cortical dysfunction. Brain magnetic resonance imaging revealed no acute vascular lesion but an old brain insult with encephalomalacia change over the right cerebral hemisphere (Fig. 1) and suspected arteriovenous malformation at the left occipital lobe. After consultation with a neurologist, conventional angiography was performed and showed occlusive lesions over bilaterally distal ICAs (Fig.

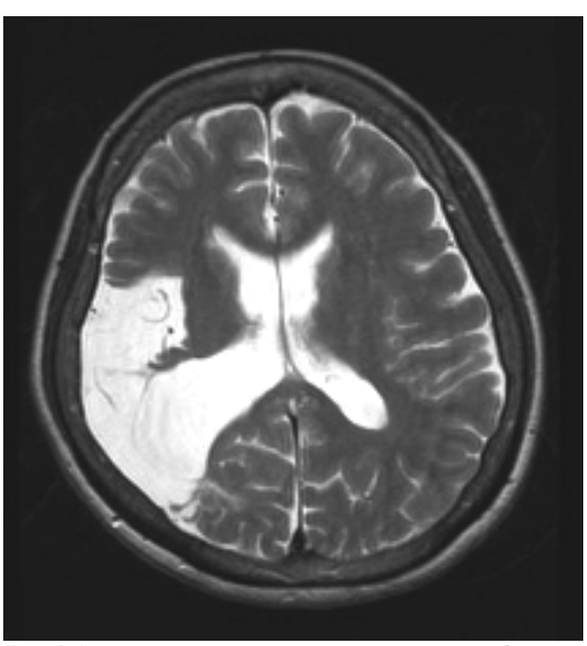

Fig. 1 Brain magnetic resonance imaging showing encephalomalacia change over the right cerebral hemisphere.

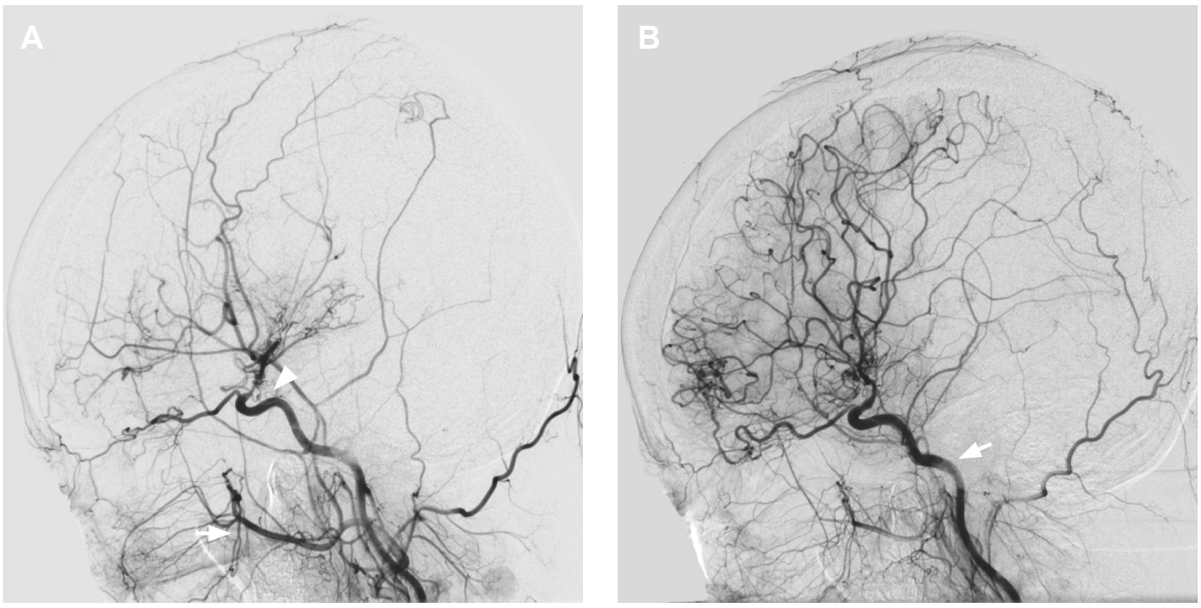

Fig. 2 (A) Occlusion of left anterior cerebral artery (ACA) (white arrow) and middle cerebral artery (MCA) (white arrowhead) with Moyamoya collaterals reconstituting MCA. (B) Severe stenosis of right distal ICA (white arrow) and occlusion of ACA, getting collateral perfusion from ophthalmic anastomosis.

2). Aspirin and rosuvastatin treatments were commenced after the decision on conservative treatment by her family.

Discussion

MMD is a rare disease and the onset tends to follow a bimodal distribution during the first or fourth decades of life. Common MMD signs and symptoms include transient ischemic attacks, strokes, seizures, headache and subsequent hemorrhage.[5] In contrast to its rare psychiatric presentation, cognitive dysfunction is more common. A previous study on 29 patients with MMD reported a broad range of cognitive impairments such as memory disturbance, executive dysfunction, and aphasia in up to two-thirds of the patients.[6] Regarding the psychiatric manifestations of MMD, depression and anxiety symptoms are more common in adults. Psychosis appears to be a more frequent manifestation in children and young adults. Mood labiality, irritability, and psychomotor agitation resembling bipolar disorder developing days to years after illness onset have also been reported, albeit less common.[7] Focusing on our patient, she presented to our Psychiatry department with atypical features (i.e., prominent cognitive deficits) and unusual treatment response (i.e., enhanced sensitivity to side effects). She has a history of major mental illness but had never received detailed neurological investigation. Despite her complaint of headache, it was very difficult to clarify the detailed associated pattern of attacks because of her cognitive deficits. Besides, common non-specific somatic complaints in patients with major mental illnesses are easily overlooked. The absence of a family history of psychiatric diseases, in combination with atypical illness features, should prompt neurological investigation. On the other side, in cases of new onset psychiatric symptoms in patients with known MMD, repeated neuroimaging examination is justified to rule out new ischemia or other vascular lesions. Psychiatric comorbidity in MMD appears to be heterogeneous with multifactorial pathogenesis. The atypical presentation in our patient may reflect an interaction between her organic brain lesion and psychiatric disorder rather than a pure contribution from the former. Our case highlights the

importance of evaluating patients with atypical presentations of psychiatric disorders for possible underlying neurological conditions.

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