

## Acute & Transient Psychosis in a Patient with Moya Moya Disease: A Rare Case Report

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**Abstract:** Moya moya disease is a rare vaso-occlusive disease of unknown etiology in which dilated collateral vessels appear hazy on angiography. Psychiatric manifestations of Moya Moya disease is uncommon. We report a case of Moya Moya disease who presented with symptoms of acute and transient psychotic disorder. This case also outlines the vitality of re-evaluating patients with acute onset, atypical or treatment-resistant psychotic symptoms for cerebrovascular pathologies.

**Keywords:** Moya moya disease, neuroimaging, psychosis, schizophrenia, acute onset

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### I. Introduction

Moya moyadisease (MMD) is a rare cerebrovascular disease of unknown etiology. It is a progressive vascular disease that leads to stenosis of the main intracranial arteries<sup>1</sup>, first described in Japan in 1957 by Takeuchi and Shimizu as a “bilaterally hypoplasia of the internal carotid arteries.”<sup>2</sup> Collateral networks of vessels, termed Moyamoyavessels, develop around the occluded arteries. Using angiographic techniques, the distal internal and proximal middle cerebral arteries are seen as narrowed or occluded, and the compensatory vascular network that develops appears as “a puff of cigarette smoke” on imaging.<sup>3</sup> Psychiatric manifestation of Moya moya disease is uncommon, few being schizophrenia, acute and transient psychosis and mania.<sup>4,7,8,9</sup> We report a case of Moya Moya disease who presented with symptoms of acute and transient psychotic disorder.

### II. Case Report

A 38 years old married female from an urban background of state of Manipur presented to the psychiatry department with irrelevant talks, muttering to self, wandering tendency, easy irritability, strong suspiciousness of being cheated and killed towards her husband & decreased sleep for a duration of 10 days which was gradually increasing in severity. On further enquiry, her sister also reported her complaints of headache and decreased appetite because of nausea and vomiting on taking solid food for last 3 days.

Mental status examination revealed uncooperativeness, decreased speech output, irritable mood and affect, delusions of persecution and infidelity. Physical examination revealed increased blood pressure (170/110 mm of Hg) with tachycardia (106 bpm). Routine blood investigations were unremarkable.

Further queries revealed with documentation that she has been diagnosed with moyamoya disease in the year 2007 when she had attack of sudden onset headache with projectile vomiting. MR Angiography then revealed intraventricular subacute hemorrhage involving right lateral ventricle with chronic arteriopathy involving the supraclinoid ICAs, proximal MCAs & ACAs with significant collateral formation, suggestive of Moya moya disease.

This time, the patient initially well responded to IM Haloperidol (5mg) in divided doses. On day 2, NCCT brain was done which showed intraventricular hemorrhage. She was then transferred under neurosurgery unit.

### III. Discussion

Moyamoya disease, although most common in Japan, few cases have been reported elsewhere, including North America, Europe and India.<sup>5</sup> Females being more commonly affected than male with ratio of 2.8:1, the disease is bimodal in distribution commonly affecting on first and fourth decade of life.<sup>6</sup> Neuropsychiatric manifestations of moyamoya disease are uncommon. Previous case reports have described psychotic association with moyamoya disease.<sup>4,7,8,9</sup> Little is known about the natural history of the disorder in neurologically asymptomatic adult patients, and the relationship between psychiatric or behavioral symptoms and Moyamoya disease is unclear.

Lubman et al (2003) reported a case of 23-year-old Vietnamese male with a 2-year history of Schizophrenia with prominent negative symptoms & executive dysfunctions. Neuroimaging revealed a prominent vascular flow void affecting the middle and anterior cerebral arteries consistent with Moyamoya disease.<sup>4</sup> Klasen et al. (1999) described a 12-year old boy who presented with an acute transient psychosis and was subsequently diagnosed with Moyamoya disease. His symptoms were precipitated by heavy physical exertion at school with no significant physical and neurological abnormalities.<sup>7</sup> Our case also presented with acute onset psychotic symptoms without any other co-morbid physical/neurological abnormalities. McDade (1991) reported a 19-year old Asian man who presented with a schizophrenia-like psychosis twelve years after diagnosis of childhood Moyamoya disease. Physical examination revealed mild right UMN signs and clumsy gait. Loss of cortex in the left temporoparietal and occipital lobes were reported in CT scan brain.<sup>8</sup> In our case too, psychotic illness appeared 11 years after she was diagnosed with Moya moya disease in 2007 with near similar neuroimaging findings. In all the patients psychotic symptoms rapidly settled on neuroleptics, similar to our case.

Like others, our case report also strictly suggests the importance of excluding organicity in patients with atypical or acute onset psychoses. It remains of utmost importance that this group of patients should have regular follow-ups for their presentations, as the psychotic illness may lead to their physical symptoms and signs being easily overlooked or falsely attributed. Because of the progressive nature of illness, it is crucial to diagnose moyamoya disease early and offer surgical management.

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