

# Presentation with Recurrent Intractable Headache: A Patient with Moyamoya Syndrome—Case Report

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## Abstract

**This clinical case report represents an interesting manifestation of a neurovascular condition that can be easily overlooked by the practicing healthcare provider. In the United States, a Hispanic patient of non-Asian descent presented with atypical symptoms of intractable headache and nausea with no evidence of neurologic deficits. Further diagnostic work-up was performed as the patient was not responding to traditional analgesic medication administration. Ultimately, cerebral angiogram revealed vascular occlusion with collateral circulation consistent with moyamoya syndrome. Discussion of the challenges and available clinical guidance for healthcare professionals dealing with patients presenting with intractable headache are presented in this report.**

Key words: moyamoya, headache, magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA)

## Introduction

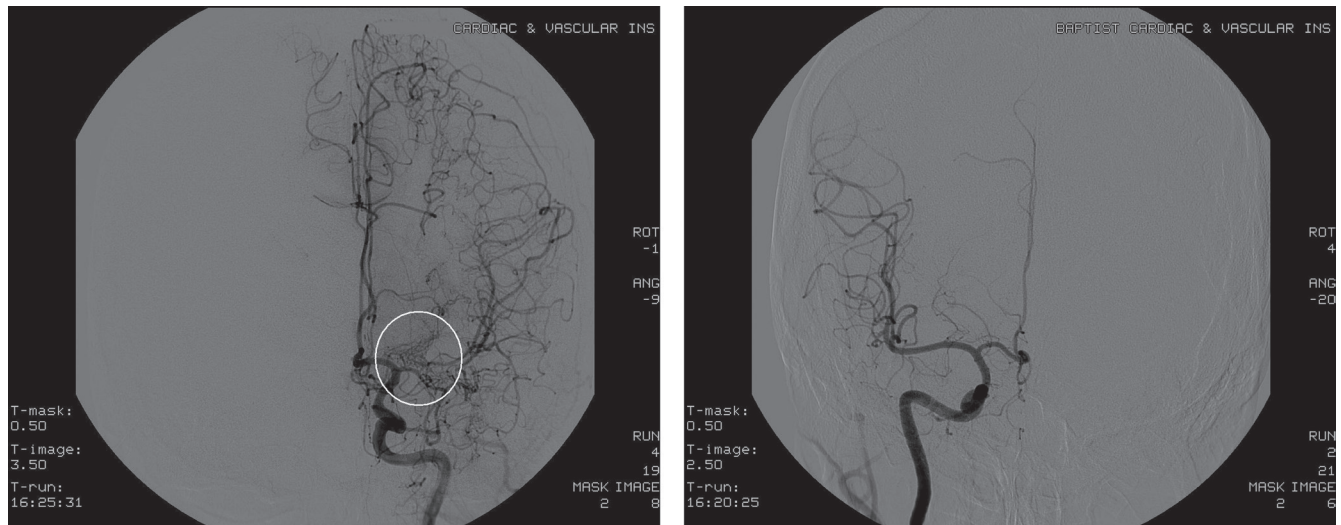
Moyamoya is a rare chronic progressive cerebrovascular disease characterized by bilateral stenosis of the distal branches of the internal carotid arteries, and the consequential compensatory development of vasculature for collateral circulation in the circle of Willis. The disorder was initially described in 1957 in Japan where the term “moyamoya,” in Japanese literally means “puffy, obscure, or hazy”.<sup>1</sup> This term was used to name the disease as it described the finely intertwined system of minute collateral vessels visible in angiograms.<sup>2</sup> Furthermore, moyamoya has approximately 10% occurrence rate in families, therefore suggesting a genetic disease etiology, although the exact causes are unknown.<sup>2,3</sup> Although variable, the primary clinical manifestations include transient ischemic attack, ischemic stroke, hemorrhagic stroke, and epilepsy, with the reoccurrence rate of each event differing by patient.<sup>2,4</sup> Repeated events of ischemic stroke or hemorrhage have also been documented with cognitive and neurological decline.<sup>5</sup>

Moyamoya disease occurs chiefly in Asian populations although cases have been observed in Caucasians, African-

Americans, and Hispanics.<sup>3</sup> Information about overall demographics for patients with moyamoya phenomena outside of Asian population is more limited.<sup>6</sup> A 2003 nationwide Japanese survey found the prevalence and incidence rates to be 6.03 and 0.54 per 100,000 population, respectively.<sup>5</sup> While data on disease rates in the United States are limited, a 2005 survey conducted in Washington and California found an incidence of 0.086 per 100,000 population, with ethnic-specific incidences of 0.28 for Asian-Americans, 0.13 for African Americans, 0.06 for whites, and 0.03 for Hispanics.<sup>6</sup> In the present clinical observation, we describe the unusual presentation of a Hispanic patient admitted to a hospital in the United States for intractable headache without a past medical history of cardiovascular or significant neurological disease.

## Objective

The objective of this clinical case report is to review the presentation, clinical diagnosis, and the treatment modalities used for moyamoya syndrome in the hospital setting. It emphasizes the challenges encountered in the diagnosis of this condition in a hospital setting, within a Hispanic patient as well as the dearth of clinical guidance available to guide a Hospitalist as to what the most appropriate



**Fig. 1** Magnetic resonance angiography (MRA) revealing occlusion of the main trunk of the left middle cerebral artery with reconstitution of the main branches through moyamoya-type collaterals (*circled*) and presentation of the right cerebral artery.

treatment is despite promising anecdotal reports about the use of antiplatelet agents.

### Case Report

A 32-year-old Hispanic female with a past medical history of migraine headaches recurring in a cyclical fashion, presented to a hospital in the United States with headache, nausea, and vomiting. She usually has migraines related to her menses; however the patient was not menstruating and was mid-cycle at time of presentation. Over the past several months prior to admission, the patient's headache severity increased in frequency and intensity. On physical examination, she was neurologically intact and no deficits were present. She was admitted to the hospital for intractable headache. Her original computed tomography (CT) scan of the head was unremarkable. Magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA) with digital subtraction angiography (DSA) of the head revealed abnormality in caliber of the left middle cerebral artery, suggestive of occlusive disease. Therefore, further work-up was warranted. Patient underwent a cerebral angiogram, which revealed occlusion of the main trunk of the left middle cerebral artery with reconstitution of the main branches through moyamoya-type collaterals from the inferior temporal branch lenticulostriate perforators and pial collaterals from the left posterior cerebral artery and anterior cerebral artery (Fig. 1).

Electroencephalogram (EEG) was negative. Two-dimensional echocardiogram (2D Echo) with bubble study showed an ejection fraction of 55–60% with normal function and no evidence of patent foramen ovale. The patient remained neurologically intact throughout her hospitalization. The

patient was screened for collagen disorders and results were negative. It was decided to begin anti-platelet therapy and she was discharged home on Aspirin (81 mg/day).

### Discussion

This clinical observation highlights two important challenges a hospitalist may encounter in the clinical presentation and management of a Hispanic patient with Moyamoya syndrome. First, this case represents an interesting manifestation of a neurovascular condition that can be easily overlooked. This patient presented with atypical symptoms of headache and nausea and no evidence of neurologic deficits. Further diagnostic work-up was performed as the patient was not responding to traditional analgesic medication administration. Ultimately, cerebral angiogram revealed vascular occlusion with collateral circulation consistent with moyamoya syndrome. There were no adverse effects reported by the patient following discharge from the hospital on the Aspirin regimen. Unfortunately, there is insufficient data in the extant literature to provide adequate guidance as to the proper short-term and long-term pharmacologic management of these patients. At the present time, treatment with antiplatelet therapy and/or anticoagulation is anecdotal among hospitalized adults with moyamoya syndrome. Further research is needed to understand the best management approach for hospitalized moyamoya patients.

### Conflicts of Interest Disclosure

The enclosed clinical observation has never been published and is not currently being reviewed by any other publication

medium. The manuscript co-authors express that they have no financial or other relationships that might lead to a conflict of interest. In addition, they have participated in the conception of this case report; and have all assisted in revising the case for important intellectual content.

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