



## A CASE REPORT OF MOYAMOYA DISEASE

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**ABSTRACT**

Moyamoya disease is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels seen on imaging, particularly cerebral angiography. The disease may develop in children and adults, but the clinical features differ. Moyamoya disease occurs predominantly in Japanese individuals but has been found in all races with varying age distributions and clinical manifestations. As a result, moyamoya disease has been underrecognized as a cause of ischemic and hemorrhagic strokes. Moyamoya disease was first described in Japan by Takeuchi and Shimizu in 1957.<sup>[1]</sup> Although the disease is most common in Japan, many subsequent cases have been reported elsewhere, including North America and Europe.<sup>[2-3]</sup> Moyamoya disease is deemed a progressive steno-occlusive disease at terminal portions of the bilateral internal carotid arteries with the development of "moyamoya vessels" as collateral channels of circulation.<sup>[4]</sup> The appearance of these small, multiple vessels at the base of the brain on catheter angiography was originally described by the Japanese term *moyamoya*, which translates to "puff of smoke."<sup>[5]</sup>

**KEYWORDS :****INTRODUCTION**

Moyamoya disease (MMD) is a progressive cerebrovascular disorder characterized by progressive stenosis of the terminal portions of the intracranial internal carotid arteries due to hypertrophy of smooth muscle in the vessel walls [6-7]. Reduced blood flow to the brain leads to the growth of collateral vasculature such as branches from small leptomeningeal vessels. Imaging of the collateral vasculature was characterized in Japan as a "puff of smoke," or moyamoya in Japanese. Incidence of MMD is the highest in Japan, where there are three cases per 100,000, and in females (2:1 ratio) [8]. MMD is now seen throughout the world with a bimodal age of onset, with children presenting around age 5 and adults presenting around age 40 [8]. The presentation in a child is typically a stroke or a transient ischemic attack, while in adults it is typically a hemorrhage. Less commonly, MMD may present with a headache or a seizure. MMD affects bilateral internal carotid arteries, whereas a unilateral presentation of the same underlying pathology is known as Moyamoya syndrome and is associated with conditions like Down syndrome, neurofibromatosis type 1, and sickle cell disease.

**CASE DESCRIPTION**

A 28-year-old young man presented to the hospital with generalized right side weakness involving right upper and lower limbs which was insidious in onset after the patient got up from the bed and was getting ready for the routine activities. The patient was disoriented and weak.

On evaluation, Patient did not have any significant past history and not similar episode in the past. No family history of stroke, seizure, or cancer. patient did not smoke, alcoholic, or drug abuser.

On physical examination, the patient was conscious and oriented. His blood pressure was 188/100 mmHg in the left arm. On neurologic examination, the cranial nerves were normal. Power was decreased (2/5) and the tone was

decreased, superficial reflexes were absent and deep tendon reflexes were exaggerated in both right upper and lower limbs. The left side neurological examination was normal, and there were no signs of sensory or cerebellar dysfunction. The complete blood cell count showed normocytic anemia with normal leukocyte and platelet counts. Coagulation profile and hypercoagulable workup was also normal. Specific tests for hypercoagulability disorders included anticardiolipin antibody, antithrombin III, Homocysteine, D-dimer, fibrinogen, lupus anticoagulant, partial thromboplastin time, protein S, prothrombin time, thrombin time, and antinuclear antibody were normal. Other investigations including a chest radiograph was unremarkable. A magnetic resonance image (MRI) of the brain with and without gadolinium contrast revealed early subacute infarcts in left basal ganglia, periventricular white matter, centrum semiovale, and just inferior to left globus pallidus showing restricted diffusion on diffusion-weighted scans. Chronic infarcts with gliotic changes were seen in right basal ganglia, periventricular white matter.

MR angiography done revealed Moderate narrowing of a proximal A1 segment of left anterior cerebral artery also seen with nonvisualization of the left middle cerebral artery and its branches. Severe stenosis also seen in right middle cerebral artery and supraclinoid segments of right internal carotid artery with paucity of peripheral branches of right middle cerebral artery.

**DISCUSSION**

Moyamoya disease is a chronic, progressive occlusion of the Circle of Willis arteries that leads to the development of characteristic collateral vessels seen on imaging, particularly cerebral angiography<sup>[9]</sup>. The incidence of MMD is the highest in Japan, but has now been reported worldwide. The disease is twice as common in females compared to males. MMD has a bimodal age of presentation. Children around 5 years of age present with symptoms of brain ischemia, often triggered by hyperventilation (e.g. transient ischemic attacks, ischemic

strokes, neurological deficits, and alterations in consciousness). Adults present in their mid-40s with recurrent headache and intracranial hemorrhage, often in the subcortical brain structures such as the basal ganglia. The hemorrhage is likely due to rupture of fragile collateral vessels or the development of cerebral aneurysms from shear stress; headaches may be due to dilated leptomeningeal collateral vessels stimulating nociceptors in the dura.

Acute management is mainly symptomatic and directed toward reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures. Revascularization procedures are currently performed to increase the perfusion to the hypoxic brain tissue. The literature supports these procedures, and the long-term favorable outcome has been reported in terms of improvement in symptoms and positive angiographic follow-ups in all age groups.<sup>[10]</sup> Hence patient was referred to the higher center for further management. On follow-up, it was found that the patient underwent direct revascularization by superficial temporal artery-MCA anastomosis [11] and had no complications in the postoperative period and was discharged.

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