

## A case report on a rare disorder: Moyamoya

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### ABSTRACT

Moyamoya is a rare disease characterized by progressive endpoint stenosis of the internal carotid arteries of the internal intracranial portions due to smooth muscle hypertrophy in the vessel walls. Here, we present the case of a 17-year-old boy who has been admitted to a tertiary care hospital in India with complaints of major headache, multiple vomiting episodes, and difficulty in seeing the right half of things from the past 20 days. The computed tomography (CT) brain, magnetic resonance imaging, and cerebral angiogram were examined for the patient. He has been diagnosed with moyamoya disease based on the evaluation. The patient was, therefore, recommended for cerebral revascularization and performed successfully then discharged with stable condition.

**Key words:** Cerebral angiography, Cerebral arteries, Internal carotid artery, Moyamoya, Surgical revascularization

Moyamoya disease is a progressive stroke characterized by gradual stenosis of the terminal sections of the internal carotid arteries because of smooth muscle hypertrophy of the vessel's walls. It is a rare disease with an incidence of 0.086 per 100,000 people reported [1]. Reduced blood flow to the brain causes collateral vasculature growth, such as small leptomeningeal vessel branches. Collateral vasculature images were characterized as a smoke puff, known as moyamoya in Japanese [2]. Vascular abnormalities are bilateral while other conditions such as neurofibromatosis, tuber sclerosis, and tuberculosis can be unilaterally present. In childhood, stroke occurs usually while in adults, subarachnoid and intracranial (IC) hemorrhage occurs [3].

### CASE REPORT

A 17-year-old male was hospitalized with complaints of severe headache in the frontal area along with several vomiting episodes and vision disturbance for 20 days. The patient was having difficulty seeing the right half of objects. The patient also had walking disturbance that was improved in 7–10 days. There was no history of fever, trauma, loss of consciousness, and surgery. There was no family history of cerebrospinal disorder, autoimmune, or ischemic heart disease.

Initially, the patient was taken to a regional hospital, and computed tomography (CT) scan, magnetic resonance imaging

(MRI), and magnetic resonance angiography of the brain were conducted. MRI brain revealed that middle cerebral artery (MCA)-posterior cerebral artery territory and bilateral supraclinoid internal carotid artery (ICA) non-visualization were shown to have acute infarcts along with proximal narrowing of bilateral cervical, petrous, and cavernous ICA (Fig. 1). Based on investigations, they were suspected of moyamoya disease and were referred to our hospital for further management.

At first, the patient was treated with aspirin and atorvastatin. Based on the above results, the patient underwent a cerebral angiogram which revealed complete occlusion of the bilateral supraclinoid anterior cerebral artery and MCA territories noted through posterior communication arteries. Based on the above-mentioned findings, the patient was diagnosed with moyamoya disease. He was suggested for surgery or brain revascularization with vascular bypass. After 10 days, surgery was performed successfully and discharged in a hemodynamically stable condition with no fresh neurological deficits. At the time of discharge, the patient was prescribed anti-platelets along with statins medications. After 1 year of follow-up, there were no fresh complaints and the patient was asymptomatic.

### DISCUSSION

Moyamoya disease represents a progressive disease of the distal carotid arteries and their major branches, characterized by the occlusion of the intima proliferation of these vessels. A distinctive finding is the formation of a collateral brain vessel network at the

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**Figure 1: Magnetic resonance imaging angiography findings revealed cerebral arteries showing puff of smoke appearance**

base of the brain with an extraordinary angiographic appearance that is described as a smoke puff [4]. Moyamoya disease has been found to be associated with conditions such as hepatitis C virus infection, cryoglobulinemia, sickle cell anemia, Type 1 neurofibromatosis, congenital cardiopathies, antiphospholipid syndrome, renal stenosis, and thyroiditis. However, the disease is not due to more than half of the kids seen [5,6].

This disease mainly affects the internal carotid arteries, which could lead to an ischemic or hemorrhagic stroke. The etiopathogenesis is not well understood but is believed to be multifactorial. Genetic factors have been involved; the genetic background is often mentioned because the illness is found among family members in 10–15% of cases. However, the patient did not have a family history in our case.

It was found, in particular, that the walls of the moyamoya affected arteries are representative of the disease with the proliferation of smooth muscle cells and of tissue deposition. These are anterior arteries, middle brain, ICAs, and posterior brain arteries. These arteries are cerebral vessels. The ICA is by far the most frequent occlusion. One hypothesis states that the disease is connected to angiogenesis caused by a fibroblast growth factor. The role of prostaglandins in the thickening of cerebral blood vessels is another postulate [7]. The reduction of the cerebral vessels appears to reject a wide range of external stimuli, injuries, and genetic defects from the brain blood vessels.

Once the blockage process begins, it tends to continue, unless surgery is treated in spite of any known health management [7]. In both children and adults, the clinical characteristics of moyamoya disease differ. No history of subarachnoid or intraparenchymal bleeding is present in adults, but in children, there has been a repeated transient ischemic attack or cerebral artery infarction. Classical disease symptoms are monoparesis, hemiparesis,

dysarthria, aphasia, headache, seizures, and unintentional movement [8].

Many of the conditions that were having similar imaging appearances to moyamoya disease were atherosclerosis, cranial radiotherapy, type 1 neurofibromatosis, tuberous sclerosis, and infections which include tuberculous meningitis, bacterial leptomeningitis, post varicella vasculitis, CNS vasculitis, systemic lupus erythematosus, and some hematological disorders such as sickle cell disease, antiphospholipid syndrome, and aplastic anemia [9]. Cerebral angiography can confirm the diagnosis of moyamoya. Surveys show that the gold standard for the diagnosis of the disease remains digital subtraction angiography. Imaging studies such as CT angiograms and high-resonance MRI show obvious vascular changes. Solar CT of photon emission is used to demonstrate a reduced blood supply and oxygen supply to the areas of the brain involving the moyamoya. High-resolution MRI is also useful for detecting atherosclerotic plaques with moyamoya progression.

Transient ischemic attack, stroke, aneurysm, or bleeding in the brain can be caused by moyamoya disease. Cognitive and developmental disabilities can also be caused. Drugs are prescribed to reduce stroke risk or control seizures. The prevention of strokes is recommended by aspirin and other antiplatelet medicines. If the patient has a seizure disorder, calcium channel blockers can relieve headaches and anti-seizure medication should be prescribed. Revascularization is recommended if the symptoms get worse or low blood flow. There are two types of revascularizations, one being a direct bypass operation involving anastomosis of the superficial temporal artery to the mid-cerebral artery. Another is indirect bypass operation that includes encephaloduroarteriosynangiosis or encephalomyosynangiosis. The moyamoya disease is used directly or indirectly or both combinations [6,10].

Medical therapy was used in moyamoya affected patients, especially when the surgery was considered high risk or patients were relatively mild. However, there are few indications of the effectiveness of medical therapy, either short or long term. A large Japanese survey showed no major differences in the results of medically or surgically treated patients with moyamoya disease, but a recent survey revealed that 38% of 651 moyamoya diseased patients were first treated with progressive symptoms. Antiplatelet agents were used to prevent emboli in sites of arterial stenosis from microthrombi. The initial treatment processes are similar to the perioperative procedure and should include isotonic fluid intravenous hydration (usually 1.25–1.50 times the daily dose), hypotension avoidance, and supplementary oxygen administration. It is necessary to prevent hyperventilation. Serum and glucose levels should be standardized. If there is seizure activity, appropriate pharmacological agents should be treated [11]. Miyamoto *et al.* compared conservative care with IC bypass operation in 80 patients with medium medical illness and concluded that extracranial-IC bypass operation is effective against re blossomy in comparison to conservative treatment [12].

**CONCLUSION**

The moyamoya disease is rare and is the main reason for children's strokes. This case highlights early recognition and management of the disease is essential for other complications to decrease and mortality even when the patients with repetitive headaches do not relieve them from suitable medicines. Cerebral revascularization operations are the best way to reduce repeated attacks and complications and improve the health quality of life for patients with moyamoya disease.

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