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Moyamoya disease: A case report

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Abstract
Moyamoya. Disease may be a rare, progressive disease caused by blocked arteries within the basal gan glia at the bottom of the brain and form of cerebrovascular disease that begins with an obliterative vasculopathy and progresses to a compensatory proliferative vasculopathy. The term Moyamoya (MM) was coined by Suzuki and Takaku in 1969 to describe the appearance of small collaterals in the thalamus and traversing the basal ganglia due to progressive stenosis. In a young patient, Moyamoya disease is an arterial disorder that causes a stroke. This is a chronic condition characterized by bilateral stenosis and occlusion of the arteries surrounding the Willis circle, as well as prominent arterial collateral circulation. It was discovered in Japan for the first time. Classic angiographic findings of stenosis or occlusion of the circle of Willi's vessels are used to diagnose it. I describe a 9-year-old male child who was diagnosed with weakness at the age of 6-months. He had remitting slurred speech and right-sided facial droop for three weeks prior to this admission, and he had head trauma after falling from a tree. An electroencephalogram and a computed tomography (CT) of the head were used to assess him. After four days in the hospital, the child improved and he was discharged. The child's condition improved gradually as medications, physiotherapy, and speech therapy were continued, and parents were counseled about the child's condition, treatment, and management.

Keywords: Acute ischemic stroke, moyamoya disease, cerebrovascular disease, arterial disorder

Introduction
The disease Moyamoya causes constriction of certain arteries in the brain. Constriction and blood clots (thrombosis) block blood flow. To compensate for the blockage, a collateral circulation develops around the blocked vessels, but the collateral vessels are small, weak, and prone to bleeding, aneurysm, and thrombosis. These collateral vessels appear as a "puff of smoke" on conventional MR angiography. This is also the case when arterial constriction and collateral circulation are bilateral. Moyamoya syndrome is a unilateral arterial constriction that can occur when one of several conditions are present. This can also be referred to as Moyamoya secondary to the primary condition. The surgical bypass is the treatment of choice for "puff of smoke" appearance is seen on angiography [14].

Case Description
A 9-year 6-month-old male child, NCM's first born. Parents brought their child to the pediatric medical ward with complaints of weakness in the right upper limb since one day, and he was admitted to the pediatric medical ward before being transferred to the pediatric intensive care unit. He had remitting slurred speech and right-sided facial droop for three weeks prior to this admission, and he had head trauma after falling from a tree. He was discharged after undergoing a head computed tomography (CT) scan. He developed slurred speech and facial drooping as a result. His primary care physician refers him to the hospital for further evaluation and treatment. There is no significant history of surgical treatment, both current and previous.

His heart rate was 92 beats per minute, blood pressure was 110/72, respiratory rate was 20 beats per minute, temperature was 98 degrees Fahrenheit, and pulse oxygenation was 99 percent on room air during physical examination. On neurological examination, he had difficulty finding words and repeating them, as well as Acute Ischemic Stroke, but his comprehension was unaffected. He also had an upper motor neuron type right-sided central facial droop. His muscle strength was normal, and he had full sensation in all of his limbs.

The results of the blood tests revealed a normal total count WBC 8,500 Cells/cumm, Neutrophils 77.3%, Lymphocytes, 1.7%, Eosinophilia, 1.6%, Monocytes, 3.7%, Basophils 0.3%, Red blood cells 3.35 million/cumm, Haemo globin 12.7 gms %, Platelet count-1,99,000 lakhs/cumm, Bleeding time 2 mts 30 secs, Sodium-136 mEq/L, Potassium 4.1 mEq/L, Phosphorus 2.5 mg/dl, Total protein 7.1g/dl, Albumin 4.1g/dl, Sr.

Globulin 3g/dl, Sr. bilirubin0.3g/dl, decreased Clotting time-5 mts 30 secs, Calcium-6.0 mg/dl, Magnesium 1.3 mg/dl, increased Chloride-107 mEq/L, EEG and CT brain scans were used to rule out the cause. Bursts of sharp waves and spikes were seen on the electroencephalogram from the bilateral front parietal region, indicating generalized epileptic form discharge. As described, a CT scan of the brain revealed features that were likely to represent an acute infarct involving the left front to the parietal region.

There was no evidence of a hemorrhage or a space–occupying lesion. If clinically indicated, an MRI brain scan is recommended for further evaluation. The child was diagnosed with 'Acute Ischemic Stroke, Moyamoya Disease' based on the EEG and CT brain reports.

Treatment

After consulting with a pediatrician, the child was advised to keep taking the Tab. Tab. 75 mg aspirin p/o OD 500 mg Citicoline BD, Inj. 500 mg Methyl IV OD, Inj. 50 mL mannitol in 30 mints with 58 mL NS, Inj. Parents were counseled about the child's condition, treatment, and management using sodium valproate 230 mg IV, as well as physiotherapy and speech therapy. After four days in the hospital, the child improved. He remained neurologically stable throughout his stay and was discharged with outpatient follow-up. He also had physical therapy for her symptoms, and on follow-up, he showed significant clinical improvement**.**

Nursing care of the child

- Monitored neurological response to activities as it can change with changes in positioning or movement •Planned nursing care activities to minimize an increase in ICP by assessing neurological status every 2 hours.
- Possible measures to prevent flexion of the neck, shoulder, hip, or knee in order to avoid blood flow obstruction
- Checked respiratory status to see if there were any changes in neurological status.
- Muscle strength and range of motion were assessed and documented.
- Determined the extent of the problem and planned appropriate interventions by evaluating the positioning ability.
- Determined the extent of the problem and planned appropriate interventions by evaluating the positioning ability.
- Used pillows to maintain proper position or alignment in order to avoid contractures.
- Determined the type of communication deficits present and planned appropriate interventions
- Used verbal prompts and reminders to encourage people to speak.
- Facilitated family communication and realistic planning to meet the needs with minimal disruption
- provided accurate information to family members regarding the condition of the patient for better understanding and coping

Discussion

Moyamoya. disease may be a rare, progressive
disease caused by blocked arteries within the basal ganglia a t the bottom of the brain This condition is most common in children, but it can also affect adults. Blood clots, strokes, and transient ischemic attacks (TIAs) are more common in affected people, and they're often accompanied by variety of neuro muscular abnormalities muscular weakness, or paralysis seizures, deficit in consciousness, speech, sensory and cognitive, involuntary movements, and vision .some neurological abnormalities (brain tumors, tuberculosis meningitis, sickle cell disease) also associated with Moyamoya syndrome [11, 14].

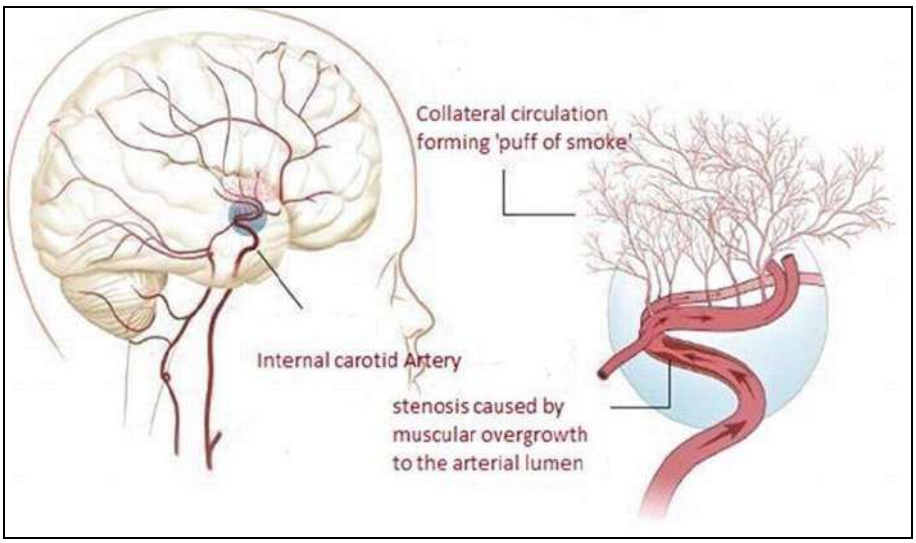


Fig 1: Pathophysiology of Moyamoya disease

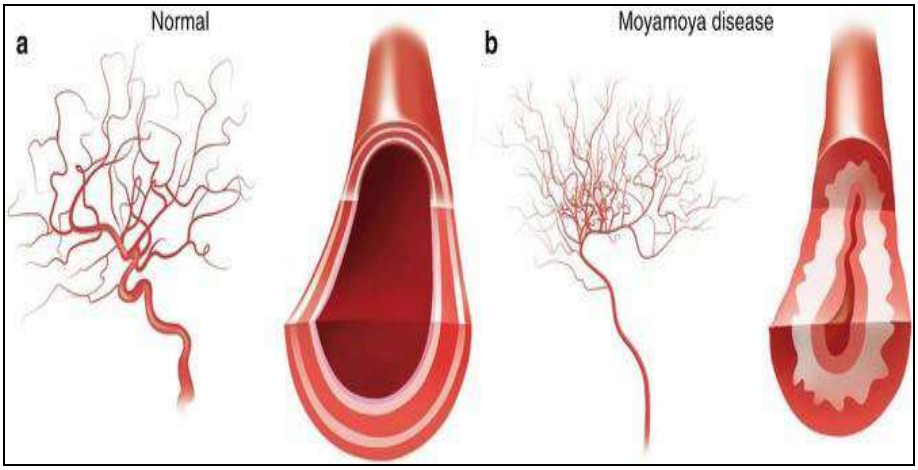


Fig 2: Normal artery and blocked artery of the brain.

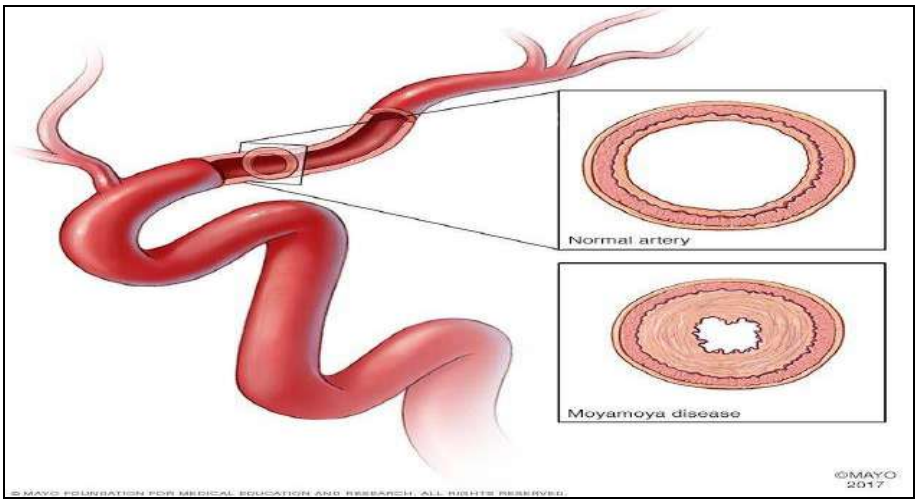


Fig 3: Narrowing of the artery due to blood clot in moyamoya disease

Etiology: Moyamoya disease has no known cause. Some patients with certain chronic diseases that alter or damage blood vessels in the brain develop Moyamoya syndrome. Sickle cell anemia, trisomy 21, X-rays of the skull, cardiac surgery, and chemotherapy are all possible causes. It is a secondary disorder that can occur in the presence of infections, vasculitis, and haematological issues. Autoimmune diseases Disorders of the connective tissue metabolic diseases, Chromosome disorders, inherited conditions, and cerebral vasculitis [1, 14].

Clinical features: Adult patients with MMD are experience intracranial haemorrhage. Adults and children can both experience seizures. Based on the aetiology of symptoms can be divided into two groups: those caused by cerebral ischemia (stroke, transient ischemic attack (TIA), and seizures), and those caused by the growth of collateral vessels that compensate for the ischemia (i.e. haemorrhage and headache). Moyamoya is most commonly associated with TIA or ischemic stroke in children. Hyperventilation caused by crying, playing a wind instrument, or eating hot noodles is a common trigger for attacks. Hyperventilation causes a reduction in carbon dioxide, which causes cerebral vasoconstriction and worsens cerebral hypo perfusion. Intra ventricular hemorrhage is common due to the close proximity of the primary site of intra cerebral hemorrhage [3].

Diagnostic Evaluation

Magnetic resonance imaging (MRI): Io find out the Hemorrhages and/or strokes in the brain parenchyma are usually detected with an MRI [4].

- Magnetic Resonance Angiography (MRA): the development of collaterals around the steno-occlusive lesions.
- Cerebral angiography: provides the size and severity of the narrowing.
- Trans cranial Doppler: Monitoring cerebral hemo dynamics,
- Electroencephalography (EEG) & CT scan: Patients who present with seizures require

It demonstrates an increase in oxygen fraction extraction, a decrease in cerebral blood flow with a posterior cerebral flow distribution, and a decreased cerebrovascular reactivity to carbon dioxide [7, 14].

There is currently no cure for Moyamoya disease. The goal of both medical and surgical treatments is to improve cerebral blood flow. Children with Moyamoya disease require a variety of conservative management and rehabilitation therapies after the initial diagnosis and treatment of stroke or TIA.

The goal of conservative management is to maintain cerebral blood flow and prevent further strokes. Patients with Moyamoya disease have traditionally been given aspirin to prevent further strokes. The usual dose ranges

from 50 to 100 mg. Symptomatic treatments with analgesics and antiepileptic medications are usually used to treat headaches and seizures. For most children who have had a stroke, rehabilitation services such as physical therapy, occupational therapy, or speech therapy is an important part of their treatment.

Paralysis of one side of the body or loss of speaking abilities, require immediate treatment. The immediate goal of treatment will be to keep the stroke from getting worse. Surgical Treatment [10, 14].

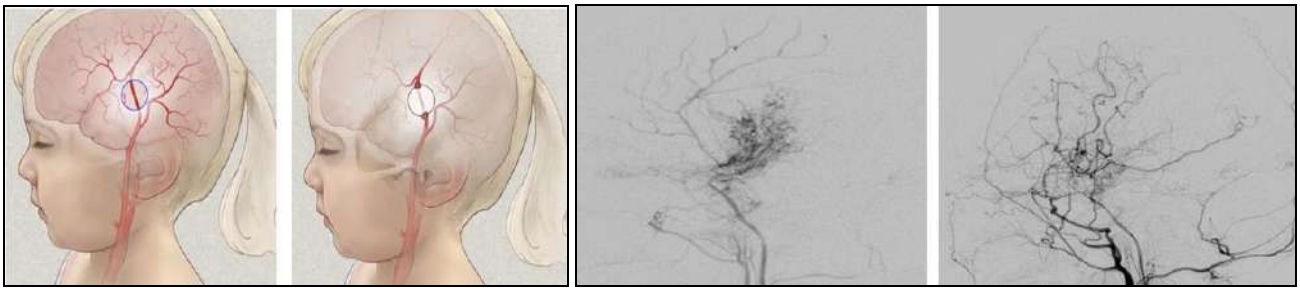
Surgical Treatment

The surgical procedure revascularization is used to treat Moyamoya disease. This procedure improves blood flow and lowers the risk of stroke by repairing narrowed arteries. Moyamoya is a progressive disease that leads to multiple strokes and mental decline if it is not treated. In perfusion studies, apparent cerebral ischemia, reduced regional cerebral blood flow, and decreased cerebral vascular reserve are the main indications for surgical revascularization. Surgery is more beneficial for children because MMD in children usually progresses quickly.

Indirect revascularization

• **Direct revascularization**: The improvement in cerebral blood flow is immediately noticeable after surgery[8]. To widen a narrowed artery, minimally invasive procedures such as angioplasty and stenting may be used in some cases [10, 13].

Surgical procedure for children with Moyamoya disease: An indirect bypass is a common treatment for children with Moyamoya disease. During this procedure, a surgeon creates a new blood delivery system for the brain. The surgeon replaces the notched portion of the skull and places the loose section of the artery directly onto the brain. The notches allow the artery to pass through the skull on either side, from the scalp to the brain (image A: right).



Left A Right B

The formation of new vessels takes months. Around 12 months after surgery, the neurosurgeon will perform an angiogram to evaluate the surgery's outcome. Image B depicts a patient with Moyamoya disease's blood delivery system before (left) and after (right) indirect bypass surgery, which resulted in the formation of new blood [11, 12, 14].

Follow-up care: After being admitted to the hospital and having surgery, most children will return within a few weeks to check on their progress. The majority of children with Moyamoya disease who receive a combination of medical and surgical treatment have a low risk of recurrent stroke. Surgery can be very effective in delivering blood to the brain and improving [11].

Prognosis: Over the course of five years, two-thirds of patients with Moyamoya disease progress asymptomatically with poor outcomes. The occlusive process continues to progress regardless of symptom severity, ongoing treatment, age, sex, disease type, or location. Surgical revascularization performed early on has a better prognosis [1].

Complications: Ischemic stroke during surgery, Ischemic stroke after surgery with a permanent neurologic deficit, Stroke with haemorrhage (0.7% -8%) Epidural hematoma (4.8% in children) after surgery after direct vascularisation, hyper perfusion syndrome develops (21.5% -50%), Scalp issues, most notably scalp ischemia (17.6% - 21.4%) [8, 9].

Conclusion

Moyamoya disease (MMD) is characterised by stenosis or occlusion of the intracranial part of the bilateral internal carotid arteries, as well as abnormal vascular collateral networks at the base of the brain. Patients with Moyamoya disease who are not treated often experience cognitive and neurologic decline as a result of repeated ischemic strokes or haemorrhages.

Patients who are diagnosed early and treated with surgical intervention as soon as possible can expect to live a normal life expectancy [13].

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