



## **A 10 Years Old Child Presented with Hemiparesis and Moyamoya Disease – A Case Report**

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### **Authors' contributions**

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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**Case Report**

### **ABSTRACT**

**Introduction:** In Moyamoya disease there is long standing, continuous developing occlusion of the arteries of Circle of Willis which leads to development of typical collateral vessels seen on cerebral angiogram. Moyamoya Disease was diagnosed on performing the MR Angiography. Child with Moyamoya disease presents with recurrent episodes of sudden hemiplegia that might occur at alternative sides. The disease is progressive and dynamic in children. Acute management of this disease is symptomatic. MR Angiography clinches the diagnosis and neurosurgical intervention with physiotherapy which may be required as a part of definitive management. Objectives: Our case brings forth the significance of considering Moyamoya Disease to be classic etiology for acute ischemic strokes in children.

**Case Report:** We report a rare case of 10 year old female child who was admitted at our medical center for the complaints of transient ischemic attack & left side hemiparesis with left facial palsy. In respect to our case, surgical intervention was not considered due to having satisfactory response on the commencing medical management.

**Conclusion:** Our case brings forth the importance of considering MMD to be classic etiology for acute ischemic strokes in children .MMD is main diagnosis when we evaluate the children with cerebrovascular events. To asses this non operative MR angiography is investigation of choice. For favourable outcome in children with Moyamoya disease the key is early diagnosis and management.

*Keywords: Moyamoya disease; stroke; MR angiography; facial palsy; puff of smoke.*

## 1. INTRODUCTION

Moyamoya disease is rare cerebrovascular condition [1]. According to Japanese Moyamoya imports to a 'hazy puff of smoke'. It is a condition characterized by progressive occlusion of cerebral vessels that is Internal carotid arteries and or the proximal portions of anterior and medial cerebral arteries [2]. According to the Ministry of Health and Welfare Japan there are mainly four types of MMD that is ischemic , hemorrhagic ,epileptic and other .Among this types ischemic is commonest in childhood , whereas in adults hemorrhagic type is most commonly observed .MMD is found to be more common in females then males worldwide [3]. According to the studies MMD is found to have some association with chromosome 17 [4]. MMD has many distinctive presenting features which includes two extremes of age groups that is at 5 and at 40 years [5]. MMD is a composite, continuously developing, multifactorial cerebrovascular occlusive condition of unrevealed etiology. Thus, it is significant to consider MMD as distinctive diagnosis and a classic etiology of stroke in young individuals [6]. So we describe a case report of a 10 year old girl who came with typical features of stroke and on investigation found to have a rare cerebrovascular disease called Moyamoya Disease [7].

## 2. CASE REPORT

A 10 year old girl child was brought to the medical Centre with chief complaints of weakness on the left side of the body, deviation of the angle of mouth towards opposite side and inability to hold things in left hand since 1 month of age .Patient also had a complain of weakness in the lower limb .Weakness was sudden in onset, observed during sleep .Patient had a history of head injury while playing 10-12 days back, with no loss of consciousness or any other feature. Patient had a complain of headache since 4 days which was intermittent in nature .There is no past medical history. There is positive family history of sickle cell anemia .There is no history of seizure.

On examination the vitals were stable, routine investigations were normal, systemic examination showed reduce muscle power in the left side .On assessing on Modified Ashworth Scale patient had grade 3 spasticity in left upper limb and grade 2 spasticity in the lower limbs.

Patient was initially started on painkiller antibiotics and physiotherapy but on performing MRI Brain Angiography a different perspective came to recognition that were the angiography finding which were suggestive of Moyamoya Disease with small infarct of right parietal lobe , following it a four limb Digital Subtraction Angiography was suggested which revealed a narrowing of right ICA supraclenoid segment and Narrowing of clinoid and Left ICA cavernous segment which were also suggestive of Moyamoya Disease .

### 2.1 Diagnostic Assesment

A case of Moya-Moya disease confirmed by cerebral angiography. MR angiography very accurately demonstrates the vascular obstruction. We get predictable narrowing of blood vessels called as stenosis or occlusion symmetrically at concluding portion of Moyamoya vessels and internal carotid artery at the base of the brain, on performing MR Angiography. Total Six phases have been reported according to angiographic finding, from Stage 1, which divulge a stenosis of the carotid branches, to Stage 6, in which the moyamoya vessels vanish and collateral circulation is produced exclusively from the external carotid arteries.

On following it a four limb Digital Subtraction Angiography was suggested which revealed a narrowing of right ICA, supraclenoid segment and narrowing of cavernous and clenoid segment of left ICA which were also suggestive of Moyamoya Disease.

### 2.2 Therapeutic Intereventions

Therapeutic measures are found to be effective to reduce the complications and improve

outcomes of patient. halting the progression of arteriopathy and treating the symptoms is focal point of treatment. In acute and symptomatic chronic phase patients as per the guidelines, it's suggested to the use of antiplatelet medications excluding in those who present with haemorrhage. For treating MMD the timing of revascularisation and surgical indications have not been established well. In order to prevent recurrent attacks of stroke and to improve the brain perfusion revascularisation surgeries are usually done. Depending on the disease presentation, revascularization surgeries should be targeted according to every individual's condition [8].

In respect to our case, surgical intervention was not considered due to having satisfactory response on the commencing medical management [9].

Physiotherapy management includes ROM exercises for elbow, shoulder, and wrist. Neuro-facilitatory techniques including Berta & Karel Bobath's approach, Rood approach with Proprioceptive Neuromuscular facilitation techniques. Functional Electrical Stimulation is also used which helps to enhance the upper extremity motor recovery. To normalize the tone stretching, passive manipulations, weight bearing, cryotherapy, contraction of antagonists to spastic muscles & splinting are given.

### 2.3 Follow up Outcomes

Since Moyamoya disease is a continuously developing condition so it do not ameliorate without proper management. While Moyamoya itself is not treatable, to provide a good long term prognosis with significant stroke risk reduction surgery is essential which provides an alternative blood flow to the brain. Death rates from moyamoya disease are estimated to be 4.3% in children and 10% in adults. In order to reduce the risk of subsequent cerebrovascular conditions and to prevent worse outcome close follow up for long time is pivotal.

### 3. DISCUSSION

Moyamoya disease is defined as an atypical cerebrovascular condition which is depicted by continuously developing occlusion or stenosis of the concluding part of the internal carotid artery and its main forks. The condition is correlated with Moyamoya vessels are widened, easily

breakable supporting vessels at the base of the brain.

Although being an infrequent condition, case studies reveal that MMD is predominantly seen in countries of East Asia with an approximate incidence of 3.2–10.5 per 1,000,000 and approx. yearly incidence of 0.43–0.94 per 100,000. Prevalence of Moyamoya disease is increasing with recent advances in neuro-radiological diagnostic modalities.

According to Case studies, females are more frequently affected than males. Patients with symptoms evolving have a better prognosis than those who present with static symptoms (which probably indicate a completed stroke). The primary presenting symptoms are cerebrovascular events and are in corresponds to both the stenosis or narrowing of the ICAs and their main forks. Recent studies shows that, if MMD is diagnosed within first year, the chance of frequent stroke was estimated to be 18% but reduced to 5% per every coming year.

Hence it's highly suggested to consider Moyamoya disease as a distinctive condition in many countries, specifically in young individuals who has features of continuously developing cerebrovascular condition.

### 4. CONCLUSION

Our case brings forth the importance of considering MMD to be classic etiology for acute ischemic strokes in children. To asses this non operative MR angiography is investigation of choice. For favourable outcome in children with Moyamoya disease the key is early diagnosis and management.

### CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

### ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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