Moya Moya Disease: A Case Report

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ABSTRACT

A chronic, occlusive cerebrovascular illness known as Moya Moya, sometimes known as "hazy puff of smoke," involves bilateral stenosis or occlusion of the proximal sections of the anterior cerebral arteries and Middle Cerebral Arteries (MCAs) as well as the terminal portion of the Internal Carotid Arteries (ICAs). The internal carotid artery becomes progressively occluded and stenotic in Moya Moya, an idiopathic illness. A 34 year old woman with a known case of Moya Moya disease has been admitted to the hospital with the same complaints of weakness in the right and lower limbs. CT Angiography showed a significant narrowing of bilateral internal carotid favoring multivessel intracranial vascular disease and right hemiparesis. She had undergone STA MCA surgery previously in June 2021. Now as she admits with the similar complaints she was posted again for STA MCA surgery in May 2022 where she showed a positive outcome post surgery. Moya Moya disease management is still not well-defined and is based on unique methods. In this patient Moya Moya disease is recurring which may make the patient prone to strokes incase of severe narrowing of arteries which may be life threatening. In order to stop more stroke episodes from occurring and to enhance outcomes, adult patients with Moya Moya disease require careful, long-term neurologic and radiologic follow-up.

Keywords: Moya Moya, Internal Carotid Arteries, STA MCA Surgery, Stroke.

INTRODUCTION

A rare form of cerebrovascular illness called Moya Moya is characterized by the gradual stenosis of significant intracranial arteries and a diffuse network of basal collaterals known as Moya Moya veins. MMD's etiology is unknown.[1] The Internal Carotid Arteries (ICAs) and the proximal sections of the anterior cerebral arteries and Middle Cerebral Arteries (MCAs) are bilaterally stenosed or blocked in Moya Moya, a chronic, occlusive cerebrovascular illness. Moya Moya Disease (MMD) is divided into four categories by the Japanese Ministry of Health and Welfare: ischemic, hemorrhagic, epileptic, and other.[2] It is more common in females than in males and has a bimodal distribution. According to research, the ischemic type is more common in children than the hemorrhagic type in adults.[3] The basic pathology of Moya Moya disease mainly includes intimal fibrous hyperplasia of the intracranial arterial stenosis, irregular proliferation of the inner elastic layer, thinning of the middle layer of the vessel wall and reduction of the outer diameter of the blood vessel.[4] Deficiency or low frequency variation of Ringin protein 213 (RNF 213) P.R4828K causes mutation of this protein on exon 61 and exon 60 causes abnormality in the development of abnormal vascular network.[5] Caveolin 1is plasma membrane protein. Decreased serum levels of caveolin1 leads to decrease in the RNF213.[6] The Pathophysiology of Moya Moya disease is explained in Figure 1. Patients can experience headaches, Seizures or cognitive impairment due to ischemic cerebrovascular accidents (CVA). The majority of patients present with a hemorrhagic stroke.[7] Characteristic angiographic findings serve as the basis for the diagnosis of MMD [not specific or sensitive]. Basal collaterals must be noticeable in order to meet the current diagnostic

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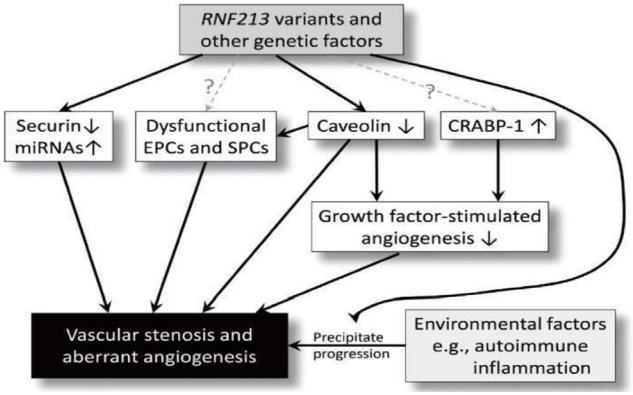

Figure 1: Pathophysiology of Moya Moya.[7]

criteria. The hallmark angiographic findings are not consistently seen in all MMD courses, and angiographic findings in MMD patients can vary depending on age at presentation and the stage of progression.[8] According to Suzuki classification of Angiographic grades Moya Moya disease is classified into 6 stages which are mentioned in the Table 1. Currently there is no evidence of drug treatment to delay or reverse progression of MMD. By exerting anti-coagulant or hemostatic actions, the present medication therapy for MMD only addresses its clinical symptoms, such as ischemia and hemorrhage.[9] Anti-platelet aggregation medications are advised for the treatment of ischemic MMD by the Japanese guidelines, however the danger of bleeding still exists.[10] To prevent the occurrence of ischemic stroke in MMD patients, surgical revascularization is preferred.[11] Surgical revascularization of MMD includes 3 types: Direct, indirect and combined revascularization.[12] Patients with MMD are usually treated with direct compared to indirect revascularization.[13]

CASE PRESENTATION

A 34-year-old woman was admitted in the department of emergency in KIMS Hospitals, Hyderabad with complaints of weakness in Right upper and lower limbs. She was a known case of Moya Moya disease, Hypothyroid, Hypertension, Diabetes. The patient was admitted in local hospital in October 2020 with similar kinds of complaints such as weakness in right upper and lower limbs, deviation of angle of mouth to left side and speech disturbances. On examination verbal commands were found to be low and the power in right upper and lower limb were 1/5 and 2/5 represented. CT angiography showed a significant narrowing of bilateral internal carotid arteries favoring multivessel intracranial vascular disease and right hemiparesis. She was then managed with Neuroprotectives, Antiplatelets, Anticoagulants, Antihyperlipidemics, Antidiabetics, Thyroid medications

Table 1: Angiographic grades according to Suzuki classification.[10]

Stages	Cerebral angiographic findings
Stage I	Narrowing of the carotid fork
Stage II	Initiation of the Moya Moya
Stage III	Intensification of the Moya Moya
Stage IV	Minimization of the Moya Moya
Stage V	Reduction of the Moya Moya
Stage VI	Disappearance of the Moya Moya

and other supportive drugs. In November 2020, she had 2 strokes and a MRI brain revealed left MCA Subcortical infarct for which treatment was given with Anticoagulants, Antiplatelets and Statins. She was on regular follow up and in February 2021 revealed she was complaining of right UL/LL spastic hemiparesis and referred for angiography. DSA performed in April 2021 provided an image of difficulty with blood flow in the brain suggestive of bilateral Moya Moya disease (left > right). She was managed medically and a plan for STA-MCA bypass was referred to. The patient underwent left STA-MCA surgery in June 2021. Follow up period was successful and was treated with supportive medicines till May 2022. Now the patient was admitted with complaints of both upper and lower right limb weakness. A plan for 2nd surgery is advised as there is a high chance of stroke attack with one phase of surgery. Second stage right STA-MCA bypass was performed in the last week of May 2022. She had shown a positive outcome Post Surgery and was managed medically with Antiepileptics, Neuroprotectives, Anticoagulants and other drugs.

DISCUSSION

A rare form of cerebrovascular illness called Moya Moya is characterized by the gradual stenosis of significant intracranial arteries and a diffuse network of basal collaterals known as Moya Moya veins. CT angiography can also be used to look for intracranial stenosis that could indicate Moya Moya, even if MRI angiography is used to confirm the diagnosis and reveal the anatomy of the arteries involved. Therefore, when MRI is not easily available and a diagnosis of cerebral occlusive vasculopathy is being evaluated, CT angiography can be examined. According to one study on asymptomatic MMD, 20% of sufferers have a silent cerebral infarction ipsilateral to the location of the Moya Moya arteries. Additionally, 40% of cases showed abnormal cerebral hemodynamics throughout the course of a 43.7-month follow-up period, including a moderate to severe loss in the cerebral perfusion reserve, increased O_2 extraction.[1] Despite the fact that some patients had stable conditions,

MMD typically progresses, with a 13.3% yearly stroke rate, and the majority of patients have repeated strokes. In ischemic MMD, surgical revascularization has been suggested to improve cerebral blood flow and stop further ischemic episodes. Theoretically, in hemorrhagic MMD, bypass surgeries are thought to stop recurrent hemorrhages by reducing the long-term hemodynamic stress on the collateral vasculature.[15] Adult MMD patients experience a chronic disease known as reduced blood flow. Therefore, it is essential to establish. Establishing healthy collateral circulation will lessen the risks of hemorrhage and ischemia. It hasn't been decided yet. whether adults with MMD symptoms should have direct or indirect surgery. The goal of this study is to share with you our conviction that mEDAS is a suitable surgical technique for treating adult MMD and that it is better for patients than EIAB.[16]

CONCLUSION

It is necessary to have a better awareness of the benefits of the various treatment modalities as well as the history of people with Moya Moya disease. Randomized clinical studies would ideally contribute to clarify the best treatment approaches. Moya Moya disease management is still not well-defined and is based on unique methods. Surgery may be dominant, especially if Moya Moya illness is identified early. The surgical revascularization for upcoming patients will probably be by improving surgical technique, perioperative care, and anesthesia. In order to stop more stroke episodes from occurring and to enhance outcomes, adult patients with Moya Moya disease require careful, long-term neurologic and radiologic follow-up.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

MMD: Moya Moya Disease; **ICA:** Internal Carotid Arteries; **MCA:** Middle Cerebral Arteries; **RNF123:** Ringin Protein 213**; CVA:** Cerebro Vascular Accident; **UL/LL:** Upper Limb/Lower Limb; **STA-MCA:** Superficial Temporal Artery Middle Artery Cerebral Artery; **MRI:** Magnetic Resonance Imaging; **MEDAS:** Modified Encephaloduroarteriosynangiosis; **EIAB:** Extracranial Intracranial Arterial Bypass.

Consent for Publication

Written informed consent was obtained from the patient's husband for publication of this case report.

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