International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Moya Moya Disease Presenting as Focal Seizures

Dr. Manjiri Naik[1], Dr. Abhay Bhosale[2], Dr. Umar Quadri[3], Dr. Sushen Ghadge[4], Dr. Siddhiraj Paramshetti[5], Dr. Shubham Patel[6], Dr. Sara Patil[7], Dr. Kahaan Shaha[8]

[1]Professor and Head, Department of Medicine, MGM Medical College and Hospital, Aurangabad

[2]Resident in Department of Medicine, MGM Medical College and Hospital, Aurangabad]

[3]Professor and Head, Department of Emergency Medicine, MGM Medical College and Hospital, Aurangabad]

[4]Assistant Professor, Department of Medicine, MGM Medical College and Hospital, Aurangabad]

[5]Resident in the Department of Medicine, MGM medical college and Hospital, Aurangabad]

[6]Resident in the Department of Medicine, MGM medical college and hospital, Aurangabad]

[7]Junior Resident in the Department of Medicine, MGM medical college and hospital, Aurangabad]

[8]Junior Resident in the Department of Medicine, MGM Medical College and Hospital, Aurangabad

Abstract: ***Moyamoya disease MMD is a rare, chronic, progressive cerebrovascular disorder characterized by stenosis or occlusion of the internal carotid arteries and their main branches. This article reports a case of an 18-year-old male presenting with focal seizures and episodic left-sided weakness, a less common manifestation of MMD. Diagnostic imaging revealed characteristic puff of smoke appearance on angiography. The patient was managed with Levetiracetam and scheduled for indirect revascularization with encephalo-duro-arterio-synangiosis EDAS. This case underscores the need for clinicians to consider MMD in young patients presenting with focal neurological deficits and seizures. Significance of the article: This article is significant because it documents a rare presentation of Moyamoya disease, providing valuable insights for clinicians in diagnosing and managing similar cases. It emphasizes the need for awareness of atypical presentations to ensure timely and appropriate treatment. Purpose of article: The purpose of this article is to report a case of Moyamoya disease presenting as focal seizures and to highlight the importance of considering this rare condition in the differential diagnosis of young patients with similar symptoms.***

Keywords: Moyamoya disease, focal seizures, cerebral ischemia, angiography, indirect revascularization

1. Introduction

Moyamoya syndrome (MMS) corresponds to the same moyamoya phenomenon but occurring in the background of either neurological or extra-neurological conditions, either inherited or acquired. A fragile network of abundant collateral vessels develops predominantly at the base of the brain in reaction to chronic brain ischemia, known as moyamoya vessels. The incidence of Moya Moya disease in the world is 0.086/1 Lakh population. Symptoms of Moyamoya disease include headache, seizures, weakness, numbness, visual abnormalities, and involuntary movements. Patients of Moya Moya Disease usually present with recurrent strokes and Generalised Tonic Clonic Seizures. However, in evidence to contrary, we report a case with complaints of focal seizures and episodic weakness in the left upper and lower limbs. He also experienced episodic slurring of speech.

2. Case Report

A 18 year old male patient came to casualty with complaints of involuntary movements of left upper limb and lower limb, which he was experiencing since 4-6 months. He was also having complaint of weakness in left upper limb and left lower limb intermittently. Approximately, he was experiencing 10-12 such episodes per day. These episodes were associated with intermittent tingling sensations in the left upper and lower limbs. He was also having intermediate episodes of slurring of speech since 1 month, approximately 4-5 episodes per day. Similarly, for the past month, he had complaints of headache in the right temporal region.
There were no complaints of bowel and bladder incontinence, fasciculations, visual disturbances, or giddiness. There was no history of syncope episode or seizure episode previously.

On general examination, he was conscious and oriented to time, place, and person, with all higher mental functions intact. His pulse was 74 bpm, blood pressure was 100/60 mmHg, and SpO2 was 99. All other systemic examinations were within normal limits.

In neurological examination his deep tendon reflexes are normal and bilateral plantars are flexors. Power in all four limbs is 5/5. Bilateral pupils are equal and reactive to light with bilateral ocular movements are with in normal limits. There was no gait abnormality & no cerebellar signs were positive.

All his lab parameters were within normal limits.

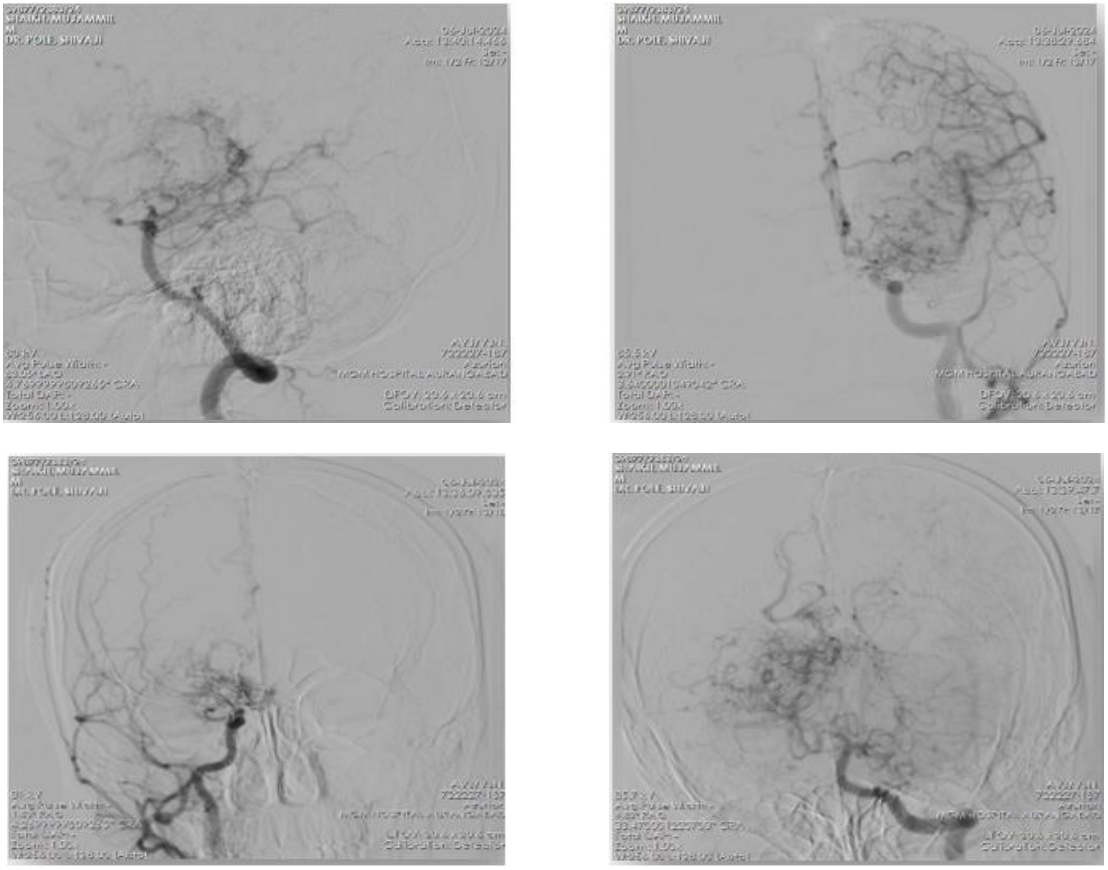
After above-described neurological examination his MRI Brain with Angiography with contrast was done which was suggestive of;

MRI BRAIN + MR ANGIO PLAIN & CONTRAST
Mild hypoplasia/ narrowing of supraclinoid right ICA with occlusion of terminal part.

Chronic occlusion right MCA and foetal origin of left PCA. Attenuated flow in bilateral ACA with hypoplastic Al segment of left ACA
Right vertebral arteries, basilar artery and right PCA appears normal.
Multiple collaterals in basal cisterns, right lenticulostriate and thalamo perforating collaterals seen giving puff of smoke appearance.

Multiple linear FLAIR hyperintensities along right cerebral sulcal spaces secondary to leptomeningeal collaterals. Features suggest p/o unilateral Moya Moya syndrome
He was then posted for Cerebral DSA, report of which was suggestive of;

Right ICA complete occlusion in distal part till the origin of right ophthalmic artery with multiple dilated vascular channels seen giving collaterals to MCA showing PUFF OF SMOKE appearance s/o Moya Moya disease.



He was staged as class 3 suzuki classification as Intensification of basal moyamoya'. On the angiographic exam, deep moyamoya vessels were intensified. MRA taken during this stage shows a "puff of smoke" appearance. The deflection of the anterior cerebral artery (ACA) and middle cerebral arteries (MCA) was noted.

He was started with tab. Levetiracetam 250 mg bd, and later posted for Indirect revascularization with encephalo-duro-arterio synangiosis (EDAS) with the supply from superficial temporal artery.

3. Conclusion

Patients of Moya Moya Disease usually present with recurrent strokes and Generalised Tonic Clonic Seizures. However in evidence to contrary, We report the case with complaints of Focal Seizures and episodic left upper limb and lower limb weakness. He also experienced episodic slurring of speech.

Hereby, we conclude that if a young patient presents with focal seizures, unilateral tingling, numbness and episodic unilateral weakness in limbs, visual disturbances, should consider Moya Moya.

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