ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 04 | April - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MOYAMOYA SYNDROME: RARE CASE IN 34 YEAR OLD MALE PATIENT IN INDIAN POPULATION – A CASE REPORT

Medicine

Dinesh Agarwal Head of the Department, Department of Medicine, Marwari Hospitals, Athgaon, Guwahati, Assam, India.

Debashish Baidya* Jr Consultant, Department of Medicine, Marwari Hospitals, Athgaon, Guwahati, Assam, India. *Corresponding Author

ABSTRACT

Moyamoya is a rare idiopathic progressive vaso-occlusive disease involving cerebral vessels like distal internalcarotid arteries on both sides, To compensate for the occlusion there is development of collateral vessels. Unilateral presentation of the condition is known as Moyamoya syndrome. Authors are presenting a case of a 34-year-old male who presented to the emergency department of Marwari Hospital and Research Centre with sudden loss of consciousness. Patient had past history of difficulty in holding objects with left hand & difficulty in walking for 3 years. The patient had undergone MRI BRAIN, diagnosed as Moyamoya disease and treated with revascularization surgery.

KEYWORDS

Moyamoya Disease, MMD, Revascularization Surgery

INTRODUCTION

Moyamoya disease (MMD) is a rare idiopathic progressive vaso-occlusive disease involving stenosis of the intracranial internal carotid arteries and their proximal branches due to hypertrophy of smooth muscle in the vessel walls[1] This leads to development of collateral vessels to compensate for the occlusion predisposing affected patients to stroke[2]Moyamoya disease is a rare disease with reported incidence of 0.086 per 100,000 population. It is seen in the people of various ethnic groups all over the world. There is bimodal age of onset seen at 5 years in paediatric age group and around 40 years in adult[3]The way of presentation also varies with age of onset and commonly present as transient ischemic attack/stroke in children while cerebral haemorrhage is the way of presentation in adult[4]MMD affects bilateral internal carotid arteries, whereas a unilateral presentation of the same underlying pathology is known as Moyamoya syndrome[5] Various risk factors for Moyamoya syndrome are female gender, Sicklecell disease, Down's Syndrome and Neurofibromatosis[6]

Case Presentation: 34 years old male patient presented with the chief complaints of sudden loss of consciousness on 10/10/2015. Patient regained consciousness after about 30 min but remained drowsy thereafter 2days and became fully conscious after that. Post-acute episode there was slurring of speech and weakness of the right side of the body, which progressed and worsened over next one week. Initially itaffected the lower limb and gradually involved the upper limb as well.

Medical history: There was no history of fever, headache, vomiting, swallowing & visual abnormalities. There was no history of head injury, no history of seizures during current illness. There was no history of involuntary passage of urine or stool.sleep and appetite was normal

PAST HISTORY: There was history of chronic headache in the past & history of seizure 2 years back for which Patient took sodium valproate in 2013 and continued it for 2 years then stopped on his own.Since last 3 years he had been having difficulty in walking, in the form of difficulty in maintaining balance with tendency to sway to left side and sensation of diminished power in left hand with difficulty in holding objects. The problem has been static and has not progressed further.

PERSONAL HISTORY: No history of addiction

Medication History: No Medication currently but history of taking anticonvulsant in the past.

PHYSICAL EXAMINATION: Patient was conscious, co-operative, and coherent, moderate build, oriented to time, place and person. Blood pressure was 110/80 mmHg, pulse rate was 78/min, all others Unremarkable.

NEUROLOGICAL EXAMINATION :

Mini Mental Status Examination score was 25.				
Speech			Normal	
Cranial Nerve examination			Normal	
Sensory System			Normal	
Motor System				
Bulk			Equal Both sides	
Power:	**Right**		**Left**	
	upper limb	3/5	Upper Limb	5/5
	Lower Limb	3/5	Lower Limb	5/5
Tone :	Diminished		Normal	
Deep Tendon Reflexes:	Biceps	1+	Biceps	2+
	Brachioradialis	1+	Brachioradialis	2+
	Triceps	1+	Triceps	2+
	Patellar	1+	Patellar	2+
	Achilles Tendon	1+	Achilles Tendon	2+
Plantar	extension		Flexion	
Spine and Cranium			Normal	
Gait :			Could not be assessed since patient unable to stand without support	

Results of pathological tests and other investigations:

Patient has under gone MRI BRAIN and findings are as follows: Sub-acute haemorrhage in the basal ganglia region extending into the periventricular and frontal areas and right side of the midbrain and pons with perilesionaledema and mass effect. Narrowing of the bilateral supralinoid internal carotid arteries, anterior and proximal middle cerebral arteries with numerous collateral vessels-**Suggesting Moyamoya Syndrome.**

Treatment plan& outcome of the treatment plan

He was advised revascularization surgery and left side indirect revascularisation and STA – MCA. Epiduro arteriomyosynargiosis was done on 18/12/2018. Postoperative period was uneventful. He did not develop any new neurological deficit and continued with the medical therapy (Antiplatelet & Anticonvulsants). Patient was discharged in a stable and ambulatory condition. Right sided revascularization is advised after 3 months

DISCUSSION

In the given case report author has presented a case of 34 years old male patient presented with Moyamoya syndrome. Moyamoya disease (MMD) is a type of chronic occlusive cerebrovascular condition involving distal internal carotid arteries on both sides and mainly anterior and middle cerebral arteries thereby leading to development of collateral vessels to compensate for the occlusion. Diagnosing MMD may be challenging but its typical presentation, underlying pathology, and appropriate angiography studies helps in diagnosis. Clinical manifestation of Moyamoya disease is variable with age of onset and usually presents with recurrent headache. The headache is migraine like in quality and refractory to medical therapies. Treatment option of

the Moyamoya disease (MMD) includes medical management and surgeries. Medical management is in the form of Low dose Aspirin as antiplatelet, anti-seizure medications, in rare instance anticoagulants, calcium channel blockers are sometimes used to help reduce headache mainly for symptomatic treatment. There is no medication available which will stop the progression of the cerebral artery narrowing and the disease will continue to progress in the vast majority of patients regardless of treatment[6]. Surgical procedures are designed to re-establish blood supply to the brain by diverting scalp blood supply to the brain surface and thereby circumventing the progressive loss of brain hemisphere blood flow. Long-term results following surgery of either type have been quite good, with long-term prevention of strokes seen in published series of both pediatric and adult patients. Daniel B. Azzam et al in 2019[3] reported a case of 35-year-old Hispanic Man with Moyamoya Disease Presented as recurrent headache while Rohit Kumar Gudepu[7] et al in 2015 reported a case Moyamoya disease in a 36 year old African American woman with repeated headache.

CONCLUSION

We conclude the given case report by highlighting the importance of considering MMD as a differential diagnosis in a patient presenting with history of chronic headache not responding to medical treatment especially in 3rd / 4th decade of life. Revascularization surgery helps to continue with day to day life by halting the progress of condition in Moyamoya disease.

Patient consent

Informed written consent for publication of clinical details was obtained from the patient.

REFERENCES

1. Miyamoto S, Yoshimoto T, Hashimoto N, et al.: Effects of extracranial-intracranial bypass for patients with hemorrhagicmoyamoya disease: results of the Japan Adult Moyamoya Trial. Stroke. 2014, 45:1415-1421
2. HUI ZHANG et al, Epidemiology, diagnosis and treatment of moyamoya disease, EXPERIMENTAL AND THERAPEUTIC MEDICINE 17: 1977-1984, 2019
3. Azzam D B, Sharma A N, Tiourin E, et al. (April 10, 2019) A Case Report of Moyamoya Disease Presenting as Headache in a 35-year-old Hispanic Man. Cureus 11(4): e4426.
4. Scott RM, Smith ER: Moyamoya disease and Moyamoya syndrome. N Engl J Med. 2009, 360:1226-1237.
5. J.A. Gosalakkal et al, Moyamoya Disease : A Review, Neurol India, 2002; 50 : 6-10
6. Sucholeiki R, Chawla J. MoyamoyaDisease.Medscape.Updated: Sep 06, 2017
7. Gudepu RK, Qureshi MA, Qureshi IA and Rao L. Case Report: A case report of Moyamoya disease in a 36 year old African American woman [v1; ref status: indexed, http://f1000r.es/4tx] F1000Research 2014, 3:297 (doi: 10.12688/f1000research.5859.1)