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A CASE REPORT OF MOYAMOYA DISEASE

General Medicine

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ABSTRACT

Moyamoya disease is a rare disease, generally idiopathic in origin. It was first described in Japan by Takeuchi. It is progressive and occlusive in nature involving mainly cerebral vessels. It generally affects bilateral distal internal carotid arteries along with anterior and middle cerebral arteries, which ultimately leads to formation of collateral circulation to compensate for this hampered circulation. It is a rare disorder with incidence of about 0.086% per 1 lakh population. Originally thought to be limited to Asian, mainly Japanese population, now it is confirmed to affect population all over the world. Incidence among female patients is almost twice, when compared to male population. It has a positive association with conditions like Sickle cell disease, Down syndrome, Neurofibromatosis, Arteriosclerosis, Irradiation and connective tissue disorders. Some cases have also shown a positive family history, with incidence ranging up to 7-10%.

KEYWORDS

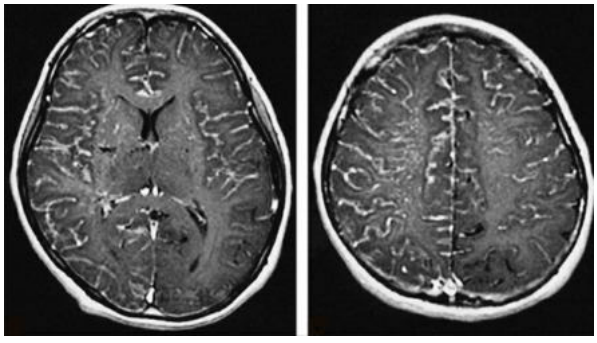
CASE REPORT

A 34 year old female presented to the emergency room with complaints of headache since 1 month and development of right sided weakness since 4 days. Headache was throbbing in nature and was not associated with any trauma. It was bilateral in nature and not associated with blurring of vision, diplopia, neck rigidity. Patient had around 4-5 episodes of headache lasting for about 1-2 hours daily and was not relieved by medications. The right sided weakness was associated with dizziness and speech difficulty. There was no fever, weight loss, sleep disturbances, vomiting, tingling, numbness or photophobia. On examination, she had a blood pressure of 138/84 mmHg and a pulse rate was 96/min, regular in rate, rhythm and volume in both upper limbs. Her Body Mass Index was 22.1 while axillary temperature was 97.5C. Patient had no significant past history and her family history was positive for Type 2 Diabetes Mellitus and Hypertension. Patient was admitted in Intensive Care Unit for proper monitoring and treatment.

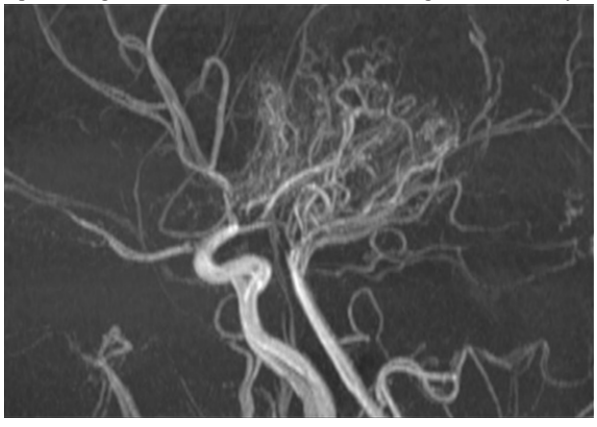
On neurological examination, pupils were bilaterally symmetrical, about 2-3 mm in diameter and reactive to light. Tone was decreased in Right upper and lower limbs, while it was normal in left extremities. Power was 1/5 in right upper limb and left lower limb, while it was 5/5 in left upper and lower limbs. Superficial reflexes were diminished, while deep tendon reflexes were exaggerated in both right and left extremities. All cranial nerve examination was normal. Basic investigations like Complete Blood Counts, Renal Function Test, Liver Function Test, Electrolytes, Thyroid Function Test were within normal limits. Coagulation study was also completely normal. Patient had intact Bowel and Bladder control with sensations present. Sensory examination of this patient was normal. There was no sign of cerebellar involvement. Electrocardiogram and Electroencephalogram were normal.

Patient was initially advised for Magnetic Resonance Imaging (MRI) with and without gadolinium contrast, following the stroke protocol. MRI revealed chronic infarcts with gliosis in right periventricular white matter along with basal ganglia. Subacute infarcts were noted in left basal ganglia, centrum semiovale and periventricular white matter. Hyperintensity was noted over right and left cerebral hemispherical sulci with collaterals development on FLAIR images, typical of "Ivy Sign''

Patient was advised for Magnetic Resonance Angiography. MR Angiography revealed narrowing in cervical segment of left internal carotid artery (70%) with severe stenosis of cavernous and supraclinoid segment of left ICA (80%) and distal flow limitation. Moderate narrowing of A1 segment of Left Anterior Cerebral Artery (ACA) territory was seen along with non visualization of left Middle Cerebral Artery (MCA). Mild narrowing was seen in right MCA with normal calibre of right MCA. Mild stenosis was noted in P1 and P2 segment of left Posterior Cerebral Artery (PCA) territory. Right PCA was normal.



FLAIR image showing Ivy sign. There is prominence of leptomeningeal collaterals. It resembles brain being covered with ivy.



MR Angiography of Intra-Cranial vessels showing classical puff of smoke appearance

Patient was immediately referred to neurosurgery department for surgical intervention and bypass procedure. Patient recovered well thereafter . Patient was later discharged on Aspirin (150mg), Clopidogrel (75mg) and Atorvastatin (40mg) once a day.

DISCUSSION

Moyamoya disease is a chronic, progressive occlusion of circle of willis, leading to formation of collaterals, which can seen on imaging. It generally presents with recurrent headaches, mimicking headaches of migraine or other common headaches. It may or may not have associated Focal Neurological Deficits. Since headaches are one of the most common feature of this disease, it should be kept in mind in

young patients presenting with recurrent headaches. These symptoms have generally a gradual course and are not responsive to routine medications. It generally causes stenosis of intracranial ICA and its proximal branches, compromising the blood supply to the anterior surface of brain. It ultimately forms collaterals near apical region of carotid artery, giving the so-called "Puffs Of Smoke" appearance on neuroimaging. This is how the disease is named, as in Japanese "Moyamoya" means "hazy like a puff of cigarette smoke". Also. Ivy sign of leptomeningeal enhancement can be seen of FLAIR images. Moyamoya disease can be diagnosed by MR Angiography, but CT Angiography can also be used. When MRI is not readily available, CT angiography can be used to detect stenosis at intracranial level. Hence, both CT scan and MRI can be used as a modality of diagnosis when occlusive vasculopathy is suspected.

Grading of Moyamoya disease based on Angiography findings:
Suzuki and Tsuchihashi's Grading system:

Grade	Definition
I	Narrowing the apex of the ICA*
II	Incipient moyamoya vessels
III	Progressive stenosis of the ICA and intensification of moyamoya vessels
IV	Development of collaterals from the ECA**
V	Intensification of ECA collaterals and reduction of moyamoya vessels
VI	ICA occlusion and disappearance of moyamoya vessels

*ICA: internal carotid artery; **ECA: external carotid artery.

Once the disease begins to develop, it is irreversible and progresses indefinitely, despite medical management. Medical management mainly involves anti-platelet drugs, but are only supportive and definitive treatment is surgery. Surgery generally involves direct and indirect revascularization procedures. Commonly used procedures are direct revascularization procedures like superficial temporal artery-middle cerebral artery bypass or middle meningeal artery- middle cerebral artery bypass. Indirect procedures like Encephalomyosynangiosis (EMS), Encephaloduroarteriosynangiosis (EDAS) and Omental Transposition can be used, but are less specific. With development in revascularization procedures, the 5 year stroke-free period in diagnosed patients with moyamoya disease is more than 90%. The mainstay of prognosis depends mainly on the time of diagnosis and surgical intervention.

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