MOYAMOYA DISEASE - A RARE CASE REPORT OF YOUNG FEMALE

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ABSTRACT
Moyamoya disease is a rare cerebrovascular disease caused by progressive stenosis of distal internal carotid artery and their major branches. Moyamoya means puffy, hazy, obscure or vague, expressing the smoky look of vascular collateral network on angiography. The dilated and fragile basal collateral circulation display “PUFF OF SMOKE” appearance, thus called Moyamoya vessels. This case highlights the importance of considering Moyamoya disease as one of the differentials while dealing with patients with recurrent headaches. Use of CT angiography as an alternative diagnostic imaging tool for diagnosing Moyamoya in cases of MRI non-availability. Cerebral Angiography remains the gold standard for these patients but which was not considered here.

KEYWORDS: Moyamoya disease, Puff of Smoke, Cerebral Angiography.

INTRODUCTION
Moyamoya disease is a rare progressive vaso-occlusive disorder of an unknown etiology. It is characterized by progressive stenosis of terminal portions of internal carotid arteries bilaterally, and the main trunks of Anterior and Middle Cerebral Artery, and is associated with collateral vessels at the base of the brain ('moyamoya' vessels).[1]

Moyamoya is a rare disease with reported incidence of 0.086 per 100,000 population. Though, originally considered to affect predominantly persons of Asian heritage it is now seen throughout the world in people of many ethnic backgrounds.[2]
In fact headache is common in Moyamoya disease and can be the first symptom especially in childhood; its characteristics and classification are largely unknown because of the uncertainty and probably multifactorial nature of the mechanism.\cite{3,4}

The symptoms include weakness of an arm, leg or both, headache, convulsions, impaired mental development, visual and speech disturbance, sensory impairment, involuntary movements or unsteady gait.\cite{5} The epileptic type of moyamoya disease is characterized mostly by focal seizures. Once the process of vascular occlusion begins, it tends to continue despite any known medical management. It leads to recurrent stroke and severe impairment of daily living functions. Moyamoya disease is rarely asymptomatic.\cite{6}

However, since the diagnostic criteria of this disease are mainly based on angiographic findings, it is recommended that the term Moyamoya ‘syndrome’ should be avoided at best.\cite{7}

**CASE REPORT**

A 21 years young female was admitted in General Medicine Department with chief complaints of headache (norco cranial throbbing type) since 4 days. History of vomiting’s since 5 days (non projectile and non bilicious) and 1 episode of seizure. Physical examination: On examination the patient was found to be conscious obeying oral commands, abdominal examination revealed no abnormalities. Blood pressure: 100/70mm Hg, CNS: B\LR Palsy +ve.

**Laboratory Investigations:** General Random Blood Sugar: 108gm/dl (80-120 mg/dl), Blood Urea Nitrogen: 34mg% (7-20mg/dl). Complete blood picture (CBP) revealed low, Haemoglobin: 10gms% (12-16gms%), RBC: 3.2Cells/µmm (4.2-5.4millioncells/µmm), WBC: 3,800Cells/µmm, (5000-10000/mm³), Platelets - reduced. Lipid profile was found to be normal.

**Differential diagnosis:** CT BRAIN revealed intra cerebral haemorrhage. MRI BRAIN impression finally revealed focal intracerebral haemorrhage noted in the left thalamus with mild perilesional edema with intraventricular extension in to CT cerebral angiogram, signs of smoke confirmed- Moyamoya disease.

**Treatment**

Initially symptomatic treatment was initiated with Inj. Mannitol 100ml/IV/TID, Tab. Naproxen 50mg/BD, Tab. Pantoprazole 40mg/OD, Inj. Levetiracetam 250mg/ IV/OD, Inj.
Sodium Valproate 100mg/IV/BD for two days. Then Tab. Atorvastatin 40mg/OD/HS, Tab. Tramadol 40mg/BD, Syp. 20% Oral glycerol 15ml/QID added and continued till 5th day.

Further treatment involved with Tab. Phenytoin 100mg/OD, Tab. Folic acid OD, Tab. Calcium D3/OD, Syp. Oral glycerol 15ml in glass of water/TID, Tab. Aspirin 75mg/OD, Tab. Pantoprazole 40mg/OD, Tab. Tramadol50mg/BD till 10th day and discharged with same medication.

DISCUSSION
Moyamoya disease is a poorly understood vascular disease primarily affecting individuals of asian descent with the largest peak at the age of 5 years. Moyamoya disease most commonly presents with seizures, stroke and episodic headache and is an important cause of cerebral stroke in children.[8]

Generally, Magnetic resonance imaging is the most common technique used in these cases. MRA proved to be a helpful diagnostic tool identifying sites of stenosis and demonstrating the collateral vessels at the base of the brain. As a non-invasive procedure, it has been described in the literature as a promising alternative to classical angiography for this arterial disease.[1,9]

When combined with MRI, it gives information about brain parenchyma and hence provides a better correlation of the symptoms with the radiological findings. However, it overestimates occlusion and underestimates presence of collaterals and can lead to higher staging of patients and lead to surgical interventions.

CONCLUSION
This case highlights the importance of considering Moyamoya disease as one of the differentials while dealing with patients with recurrent headaches who are in their third/fourth decade of life and children around 5 years old in whom headache is not relieved on routine medical treatment.

It also emphasizes on the rare presentation among Indian population and use of CT angiography as an alternative diagnostic imaging tool for diagnosing Moyamoya in cases of MRI non-availability. Cerebral Angiography remains the gold standard for these patients but which was not considered here.
REFERENCES


