



Moya Moya Disease presenting as Posterior circulation stroke

KEYWORDS

moyamoya disease , puff of smoke , posterior circulation

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ABSTRACT *Moyamoya disease is a chronic cerebral vasculopathy causing progressive occlusion of the arteries in the circle of Willis leading to the development of characteristic collateral vessels. It is common in Japan but an extremely rare entity in India. We report here a 21 year old female with a past history of left hemiparesis and bilateral visual loss at the age of 5 yrs, a transient ischaemic attack (TIA) at 15 years age and multiple episodes of seizures since 3 years. Magnetic resonance imaging (MRI) brain and angiography was suggestive of Moya moya disease.*

Introduction

Moyamoya disease (MMD) is a chronic occlusive cerebrovascular disorder characterized by progressive stenosis of the arteries of circle of Willis. The disorder initially involves the intracranial portion of the internal carotid arteries and progresses to involve the middle, anterior & posterior cerebral arteries. It is associated with the formation of an abnormal collateral vascular network due to compensatory dilatation of lenticulostriate & thalamostriate arteries¹. These abnormal vessels at the base of the brain were first described by the Japanese -Takuchi & Shimizu in 1957- as having a "hazy, cloudy puff of smoke" appearance which led to the term "moyamoya"².

Case Report :

A 21 year old female presented to the emergency department with generalised tonic - clonic status epilepticus. She had history of recurrent seizures for the last 3 years and was on tablet sodium valproate (200mg) twice daily. She had omitted medications on her own 2 months ago. There was history of sudden loss of vision in both the eyes and left hemiparesis at 5 years of age, diagnosed as an ischemic stroke. At 15 years of age she suffered from a transient ischemic attack presenting as mild right upper limb weakness with tingling numbness which recovered completely within four hours. There was no history or symptoms suggestive of vasculitis or systemic infection. She attended a school for the blind. Family history was not contributory.

On general physical examination she was afebrile, pulse- 92/min, BP 110/80 mm of Hg. Both carotids were normal, with no bruit. On ophthalmic examination, light perception was absent in both the eyes. Fundus examination was within normal limits, pupils were normal equally reacting to light on both sides. Examination of other cranial nerves showed no abnormality. She had left hemiparesis with power of grade 3 in upper limb and grade 4 in the lower limb. Her left leg was spastic with an extensor plantar response. Laboratory investigations revealed hemoglobin was 12.2 gm/dl, leukocyte count 5130/cumm, platelet count 2.11 lakhs, blood sugar level 82 mg/dl. Thrombophilic profile (ANA, anti dsDNA, anticardiolipin antibody, antiphospholipid antibody (APLA) and homocysteine levels) were within normal limits. Chest radiograph and electrocardiogram did not show any ab-

normality. Her conventional carotid angiography at the age of five years revealed multiple small collateral vessels (Puff of Smoke appearance) (Fig 1). MR Angiography brain showed bilateral supraclinoid carotid occlusions and bilateral Posterior cerebral artery (PCA) appear smaller in calibre with multiple small collateral channels in right parieto- occipital lobe region (Fig 2). MRI Brain FLAIR showed Gliotic changes in right temporo-parieto- occipital region and left occipital region with flow voids in right internal capsule and left caudate nucleus (Fig 3).

She was put on antiepileptic medication Tab Levetiracetam (250mg) twice daily and Tab. Ecosprin (150mg) once daily. There was no episode of seizure during her hospital stay.

We counselled the patient & parents regarding the medical, surgical treatment and outcome of the disease. We offered her an extracranial- intracranial bypass surgery which she refused.

Discussion :

Primary and secondary forms of Moyamoya disease are recognised. Primary MMD has a strong hereditary predisposition. It is common among Japanese patients. The incidence is 0.1 in 100000 per year. The gene responsible for this disorder is located on the short arm of chromosome 3. In the pediatric age group girls are more frequently affected with the peak age of onset 5 years of age³, as in our patient. Secondary forms are termed moyamoya syndromes (MMS) and have been identified in a large number of conditions such as atherosclerosis, neurofibromatosis, post radiation therapy, sickle cell disease and more recently in association with Down's syndrome¹. Our case had no such cause so she was diagnosed as a case of primary moyamoya disease. The diagnosis of MMD is based on three main criteria - 1) Stenosis or obstruction of the distal intracranial segment of the internal carotid arteries and the proximal arteries and the middle cerebral arteries 2) Abnormal vascular network (moyamoya) on the arterial side near the obstruction or stenosis, arising from lenticulostriate and thalamostriate arteries 3) Exclusion of meningitis, sickle cell disease, tumour, trauma or radiation therapy. Although unilateral presentation of MMD has been reported, bilateral involvement is considered as another criterion of diagnosis¹. The Japanese have classified

MMD into four types.

- The hemorrhagic type characterised by subarachnoid bleeding.
- The epileptic type with repeated seizures.
- The infarct type with permanent gliosis.
- The transient ischemic attack type marked by recurrent transient ischemic attacks³.

Clinical presentation of adult-onset MMD differs from childhood form. About 40%- 65% of adults present with intracranial bleed, either intraparenchymal hemorrhage with intraventricular extension or subarachnoid hemorrhage. In children only 10% present with hemorrhage while majority present with focal ischemic deficits. Suzuki and Kodama had shown that adults have less ability to form collateral vessels as compared to children.²

Our patient had infarcts in bilateral occipital and right parietal regions at the age of five years and presented to us with blindness, left hemiparesis and multiple seizure episodes. She also had a history of transient ischemic attack at the age of fifteen years. Recent MRI revealed gliotic areas in the regions of old infarct. Thus she had a mixed clinical presentation.

An unusual feature in our case was the occurrence of infarction in the vascular territory of the posterior cerebral arteries. Most infarcts have been described in the anterior circulation. Though angiographic abnormalities have been described in the posterior circulation, infarction in the territory of the PCA (posterior cerebral artery) is distinctly rare. Only 11 out of 82 cases studied by revealed PCA territory infarcts⁴. A recent study by Mugikura et al⁵ reported higher degrees of posterior cerebral artery stenosis and a higher prevalence of completed stroke in children who were diagnosed at ages < 4 years, as compared to those diagnosed at older ages

Pathological changes in MMD include intimal thickening with fibrous tissue, abnormalities of internal lamina elastica, variable lipid deposition and virtual absence of inflammatory reaction in the blood vessels¹. Though conventional angiography or DSA (Digital subtraction angiography) are investigations of choice MR angiography has become a reliable diagnostic modality for MMD. MRI not only reveals areas of infarctions but also allows direct visualisation of collateral vessels as multiple flow voids usually in the region of lentiform nuclei. MR angiography is used to confirm the diagnosis and to see the anatomy of the vessel involved⁴. In children the first line of treatment is pharmacological with antiplatelet therapy and calcium channel blockers, which can improve symptoms but do not halt disease progression⁷.

Several surgical procedures, which can be classified as direct and indirect bypass methods, have been proposed for the treatment. The direct bypass techniques that have been proposed are vein grafts and EC-IC (Extracranial- Intracranial) arterial anastomoses, Superficial Temporal Artery- Middle cerebral artery (STA-MCA) and Occipital artery - Middle cerebral artery (OA- MCA) anastomoses. The indirect techniques are as follows: 1) Encephaloduroarteriosynangiosis 2) Encephalomyosynangiosis 3) Encephalomyoarteriosynangiosis 4) use of multiple cranial bur holes 5) transplantation of omentum. Other options such as cervical carotid sympathectomy and superior cervical plexus ganglionectomy have also been proposed. Medical management had not been proven capable of controlling or improving disease⁶.

Conclusion :

We have presented this case to increase the awareness of this entity among physicians, especially post graduate students. Moyamoya disease is an extremely rare condition in India but should be considered in patients presenting with recurrent stroke and seizures. Also, posterior circulation stroke is a relatively rare occurrence with Moyamoya disease. It is crucial to offer surgical treatment to the patient early in order to reduce

the mortality and morbidity.



Fig 1 – Carotid angiography of left vertebral artery showing extensive collaterals in posterior circulation (Puff of smoke appearance).



Fig 2 : MR angiography shows bilateral supraclinoid carotid occlusions and bilateral Posterior cerebral artery (PCA) appear smaller in calibre with multiple small collateral channels in right parieto-occipital lobe region.

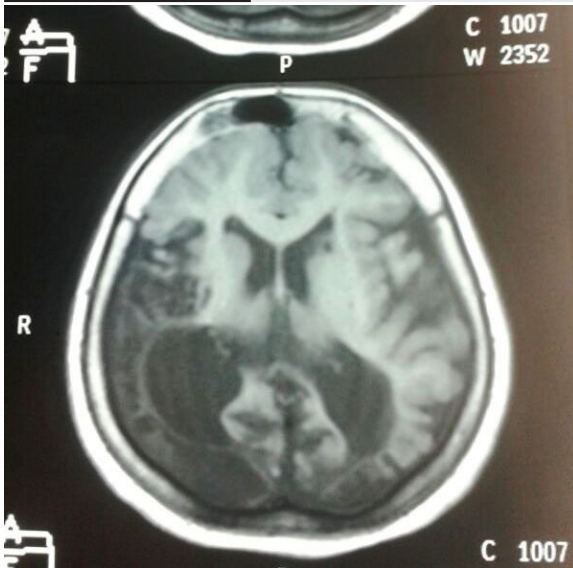


Fig 3- MRI Brain FLAIR showing Gliotic changes in right temporo-parieto- occipital region and left occipital region with flow voids in right internal capsule and left caudate nucleus.

Legend-

Fig 1 – Carotid angiography of left vertebral artery showing extensive collaterals in posterior circulation (Puff of smoke appearance).

Fig 2 - MR angiography shows bilateral supraclinoid carotid occlusions and bilateral Posterior cerebral artery (PCA) appear smaller in calibre with multiple small collateral channels in right parieto- occipital lobe region.

Fig 3- MRI Brain FLAIR showing Gliotic changes in right temporo-parieto- occipital region and left occipital region with flow voids in right internal capsule and left caudate nucleus.

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