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Case Report

Unraveling the mysteries of moyamoya: a rare case report of stroke in young

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ABSTRACT

Moyamoya disease is a unique cerebrovascular disease that is characterized by chronic progressive stenosis of distal part of internal carotid artery with consequent development of a network of collateral vessels in response to brain ischemia. It is mainly seen in individuals of Asian descent and is the most common cause of stroke in Asian children. However, it is rare in Indian subcontinent. Here we report a case of young adult who manifested with moyamoya disease, evident from acute onset ischemic stroke. The patient underwent a diagnostic cerebral angiogram that showed bilateral posterior cerebral artery stenosis with pathognomonic collateral moyamoya vessels. The patient subsequently underwent elective surgical intervention procedures to prevent further complications.

Keywords: Moyamoya, Stroke, Young

INTRODUCTION

Moyamoya, a rare cerebrovascular condition, is defined by gradual and progressive stenosis of the intracranial part of internal carotid arteries. As a result, it triggers the development of compensatory arterial collaterals. The vessels have a typical smoky appearance on the angiogram, first called 'moyamoya' a Japanese expression meaning hazy like 'puff of smoke'.

Moyamoya disease is more common in the Asian population and predominantly affects children. It has a bimodal age distribution which peaks in the first and fourth decades of life. Childhood moyamoya is characterized by ischemic manifestation while adult moyamoya presents with hemorrhagic manifestations.[1]

Although the exact cause of moyamoya disease remains unknown, the high incidence of the condition among people of East Asian descent suggests a genetic predisposition.[2]

Here we reported a case of this rare disease in a 17-year-old male with bilateral posterior cerebral artery occlusion and multiple collateral formation on MR-angiography of the brain.

CASE REPORT

A 17-year-old male presented in medical emergency with acute onset weakness of right upper and lower limb for last 12 hours which was preceded by headache and one episode of focal seizure with intact awareness.

He had a similar history of acute onset left sided weakness 4 years ago from which he recovered after 1 month with no residual neurological deficit. He has normal birth and developmental history. Family history was non-contributory.

Upon examination, vitals were within normal limits. Neurological examination revealed paucity of movement of right upper and lower limb with power of 2/5. Vision

examination revealed homonymous hemianopia. Right planter was up. Modified Rankin Score (mRS) at admission was 4. An emergent CT scan without contrast revealed hypodensity in left temporal, parietal and occipital region of brain parenchyma. Treatment of ischemic stroke was initiated with aspirin and atorvastatin 20 mg. Anti-Epileptic drugs (AED) were also administered to prevent further episodes of seizure. Thrombolysis was not done as patient was out of 4.5-hour window.

MRI brain (Figure 1) was done which revealed features suggestive of acute ischemic stroke in left hemisphere of brain.

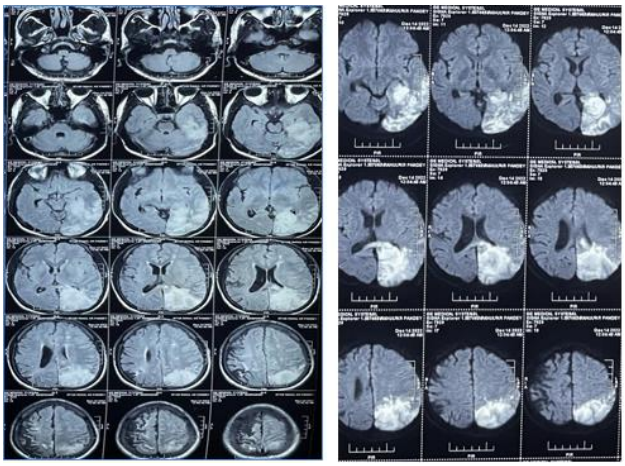

Figure 1: FLAIR showing hyperintensities in left occipital, parietal and temporal lobe with gliosis and brain parenchymal atrophy on right hemisphere. Diffusion-weighted MRI showing diffusion restriction in left occipital, temporal and parietal cortex.

MR-angiogram brain (Figure 2) revealed left posterior cerebral artery (PCA) completely occluded with diffuse bilateral net of enhancing small vessel collaterals. Right posterior cerebral was partially occluded with multiple collaterals.

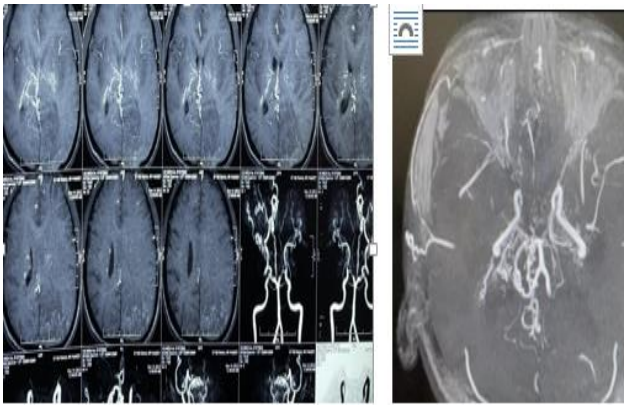

Figure 2: MR angiogram of brain showing right PCA partially occluded and left PCA completely occluded with diffuse bilateral net of enhancing small vessels denoting collaterals.

Further workup including CBC, LFT, KFT, ANA profile, beta-2-microglobin, anti-cardiolipin antibody, HIV antibody, serum complement level were all unremarkable. Trans-thoracic echocardiogram and doppler of neck vessels were normal.

Patient was discharged on day 7 of admission with mNIHSS of 5. At discharge patient was prescribed aspirin, atorvastatin and AED. On follow up, after 30 days patient showed complete recovery of neurological weakness (mRS of 0) and no episode of seizure. He had occasional episodes of headache. With further consultation in neurosurgery OPD he was planned for PCA bypass surgery.

DISCUSSION

Moyamoya is a well-established clinical and pathological entity but its exact etiologic mechanism remains unknown. There are two distinct forms in which the condition can manifest, namely Moyamoya disease (MMD) and Moyamoya syndrome (MMS). MMD refers to the idiopathic occurrence of a disease without any risk factors, whereas MMS describes the manifestation of the condition in conjunction with other risk factors like neurofibromatosis 1 (NF-1), sickle cell disease, Down syndrome, etc.[3]

Determining the prevalence of moyamoya disease poses challenges due to its rarity and the fact that it may not always be diagnosed.

Nevertheless, prevalence estimates range from 0.1 to 0.5 cases per 1,00,000 patients.[4] In the conducted surveys, the ratio of males to females was reported as 1:1.8 or 1:2.2, and around 10%-15% of the patients had a familial medical background.[5]

From a pathological standpoint, MMD exhibits features such as thickening of the fibrocellular intima, proliferation of smooth muscle cells, and increased accumulation of elastin. There is also thinning of tunica media which may result in pseudoaneurysms leading to haemorrhage.

Although traditionally thought to only affect the anterior circulation, more than half of cases actually involve the posterior circulation.[6]

MRI angiography is a diagnostic tool used to confirm a diagnosis and identify the anatomy of vessels involved in cerebral occlusive vasculopathy. In cases where MRI is not readily available, CT angiography can also be used to identify intracranial stenoses that suggest moyamoya.[7]

Based on angiography Suzuki staging was developed in 1969 which is still used today (Table 1). The involvement of the posterior circulation in moyamoya disease has not been thoroughly investigated.[8] If the PCA is involved, it primarily affects the distal portion.[9] This is because the posterior circulation acts as a collateral pathway to

maintain circulation. It is worth noting that the involvement of the posterior circulation in moyamoya disease represents an adverse prognostic factor for the disease outcome.[10] Furthermore, ischemic episodes are usually infrequent in the posterior circulation territory until the late phase of moyamoya diseas.[11] These observations underscore the importance of monitoring the progression of the disease closely, especially in cases where the posterior circulation is involved.

Table 1: Suzuki staging in moyamoya disease.

Stages	Angiographic finding
I	Narrowing of the carotid bifurcation
II	Initiation of the moyamoya: continued narrowing of the ICA; dilation of the ACA and MCA, initial moyamoya blush
III	Intensification of the moyamoya: loss of proximal ACA and MCA; leptomeningeal collateralization from the PCA; increase in moyamoya blush
IV	Minimization of the moyamoya: progressive occlusion of ICA reaching origin of PCA; reduction in moyamoya blush
V	Reduction of the moyamoya: complete loss of ICA, ACA, and MCA; increased collateral supply from ECA; further reduction in moyamoya blush
VI	Disappearance of the moyamoya: disappearance of blood supply from ICA; blood supply exclusively from ECA; disappearance of moyamoya vessels

The optimal treatment for ischemic moyamoya is unclear. Platelet antiaggregants, vasodilators, calcium channel blockers, and corticosteroids have been used with varying results. Surgical intervention is generally required for MMD treatment.

CONCLUSION

When encountering sudden focal neurological deficits in young adults, it is essential to consider less common causes of stroke, such as moyamoya disease and syndrome, due to their therapeutic and prognostic implications. Despite the variable and unpredictable nature of this condition, revascularization surgery is recognized to yield favorable outcomes, particularly if undertaken before the occurrence of irreversible neurological damage.

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