

Varied presentations of moyamoya disease in a tertiary care hospital of north-east India

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Abstract

Introduction: Moyamoya disease is a chronic progressive cerebrovascular disorder, characterized by stenosis or occlusion of bilateral internal carotid arteries (ICAs), anterior cerebral arteries (ACAs) and middle cerebral arteries (MCAs), accompanied by a collateral network of vessels formed at the base of the brain. Ischemia and intracranial hemorrhage are the common typical manifestations. However moyamoya disease has been associated with atypical presentations like headache, seizures and involuntary movements. Although frequently reported from Asian countries like Japan, China and Korea, only few studies reported on clinical manifestations of moyamoya disease from India. **Objectives:** To study the varied presentations of moyamoya disease in a tertiary care hospital of north-east India. **Material and Methods:** Relevant investigations were done to rule out other causes of moyamoya syndrome. **Results:** We report 6 cases of moyamoya disease with varied presentations from a tertiary care referral government hospital. Case 1, 2 and 6 presented with alternating hemiparesis. Case 3 had amaurosis fugax. Case 4 had history suggestive of ischemic stroke and presented with hemichorea. Case 4 had focal seizure as the only manifestation. Cases 4 and 5 notably had stenosis of posterior cerebral artery (PCA) in addition to stenosis of bilateral ICAs, ACAs and MCAs. **Conclusion:** Owing to its low incidence in India, moyamoya disease is easily overlooked as a possible diagnosis. However, because of its progressive nature, it is imperative to diagnose this disease early and offer surgical treatment to the patients.

Key Words

Cerebral infarction, moyamoya, transient ischemic attack

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Introduction

Moyamoya disease is a chronic progressive non-atherosclerotic, non-inflammatory and non-amyloid cerebrovascular disorder, defined as progressive stenosis of the intracranial vessels. The stenosis begins from the terminal bifurcation of internal carotid arteries and gradually progresses to the anterior, middle, and posterior cerebral arteries.

The clinical features of moyamoya disease differ substantially between children and adults. Most children with moyamoya

disease develop transient ischemic attack (TIA) or cerebral infarction, whereas about half of adult patients develop intracranial hemorrhage (ICH), and half develop TIA or cerebral infarction. There are two main causes of intracranial bleeding in moyamoya disease: Rupture of dilated, fragile moyamoya vessels or rupture of saccular aneurysms in the circle of Willis. Other neurological manifestations reported in moyamoya disease are headache and epilepsy.^[1,2]

Despite many advances in the past 50 years, moyamoya disease remains an enigma with many aspects of the disease remaining unsolved.^[3] We report six cases of moyamoya disease with very rare presentations from the Department of Neurology of a tertiary care referral government hospital.

Case Report

Case report 1: Alternating hemiparesis at presentation

An eight-year-old male child presented in emergency department with sudden onset left-sided hemiparesis and altered sensorium. History was suggestive of right-sided

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hemiparesis with complete recovery at the age of 5 years. On examination, the child had the following findings: Drowsy, normal vitals, normal cranial nerves, normal fundoscopy, left hemiparesis (power 3/5); exaggerated left deep tendon jerks and extensor left plantar response. The following investigations done in evaluation of stroke and were found to be normal: Complete blood count with peripheral blood smear, erythrocyte sedimentation rate (ESR), renal function tests, liver function tests, blood sugar and serum electrolytes; antistreptolysin O (ASO) titer, hepatitis B surface antigen (HBsAg), anti-hepatitis-C (Anti-HCV), venereal disease research laboratory (VDRL) and human immunodeficiency virus-1(HIV-1), 2; electrocardiogram (ECG), chest X-ray and echocardiography. MRI Brain with MRA showed narrowing of supraclinoid left ICA, ACA and MCA, supraclinoid right ICA and MCA. He was diagnosed as a patient with moyamoya disease. Unfortunately the child's sensorium deteriorated and he expired due to aspiration pneumonia.

Case report 2: Alternating hemiparesis at presentation

A ten-year-old female child presented with sudden onset right-sided hemiparesis. Past history was suggestive of left-sided hemiparesis with complete recovery at the age of 8 years. On examination, the child had the following findings: Conscious, afebrile, normal vitals, normal cranial nerves, normal fundoscopy, right hemiparesis (power 3/5), exaggerated right deep tendon jerks, and extensor right plantar response. The following investigations done in the evaluation of stroke and were found to be normal: Complete blood count with peripheral blood smear, ESR, renal function tests, liver function tests, blood sugar and serum electrolytes; ASO titer, HBsAg, Anti-HCV, VDRL and HIV-1, 2; ECG, chest X-ray and echocardiography. MRI Brain with MRA showed narrowing of bilateral supraclinoid ICA, ACA and MCA. The child was treated conservatively. Follow up at 2 months showed presence of mild residual weakness.

Case report 3: Amaurosis fugax at presentation

A twenty-year-old female had recurrent episodes of painless and transient loss of vision in right eye (for about 15 min) for one week. There was no history of weakness of limbs, headache, loss of consciousness or seizure. On examination, her vitals were normal, fundoscopy normal and there was no objective neurologic deficit. Blood investigations (including complete blood counts, ESR, liver function tests, renal function tests, blood sugar, fasting lipid profile and serum electrolytes) were normal. ECG, chest X-ray, carotid Doppler and echocardiography were normal. Electroencephalogram (EEG) was normal. MRI Brain with MRA showed bilateral narrowing of ICA, ACA and MCA.

Case report 4: Hemiparesis with hemichorea at presentation

A ten-year-old female child had history of involuntary movements for 15 days. 6 months ago, the child had sudden onset of right-sided hemiparesis, which improved but she had residual weakness. On examination, she was conscious and her vitals were normal, cranial nerves normal, fundoscopy normal. Her right hand grip was weak. She had irregular jerky involuntary movements of the left side of body. Complete blood counts with peripheral blood smear, ESR,

liver function tests, renal function tests, blood sugar, serum electrolytes, TSH and ASO titre were normal. ECG, chest X-ray and echocardiography were normal. Slit lamp examination was normal. EEG was normal. MRI Brain with MRA showed narrowing of bilateral supraclinoid ICAs, ACAs and MCAs, extensive collaterals of perforating arteries and narrowing of bilateral PCAs.

Patient was treated with oral haloperidol and sodium valproate. At the time of last follow up visit chorea have subsided with residual hemiparesis.

Case report 5: Focal seizures at presentation

A nine-year-old female had recurrent episodes of jerky movements of right side of body, lasting about 5 min, for one week. On examination, the child was conscious, vitals were normal and there was no objective neurologic finding. The child was diagnosed to have focal seizures and was investigated. Complete blood counts with peripheral blood smear, ESR, liver function tests, renal function tests, blood sugar and serum electrolytes were normal. Contrast enhanced computed tomography (CECT) Brain was normal. EEG showed paroxysmal spikes and waves discharges from left fronto-temporal leads with generalization [Figure 1]. MRI Brain with MRA showed focal narrowing in bilateral ICA in its terminal part, proximal MCA, ACA and left P1 segment of PCA with multiple collaterals. She was treated with oral levetiracetam.

Case report 6: Alternating hemi paresis at presentation

A fifteen-year-old male presented with alternating hemiparesis. On examination, the child was conscious, afebrile with normal vitals, normal cranial nerves, normal fundoscopy, right hemiparesis (power 4/5), exaggerated right deep tendon jerks, and extensor right plantar response. Complete blood count with peripheral blood smear, ESR, renal function tests, liver function tests, blood sugar and serum electrolytes; ASO titer, HBsAg, Anti-HCV, VDRL and HIV-1, 2; ECG, chest X-ray and echocardiography revealed no abnormality. MRI brain [Figure 2: Panel A] with MRA [Figure 2: Panel B] showed narrowing of bilateral supraclinoid ICA, ACA and MCA. The child was treated conservatively.

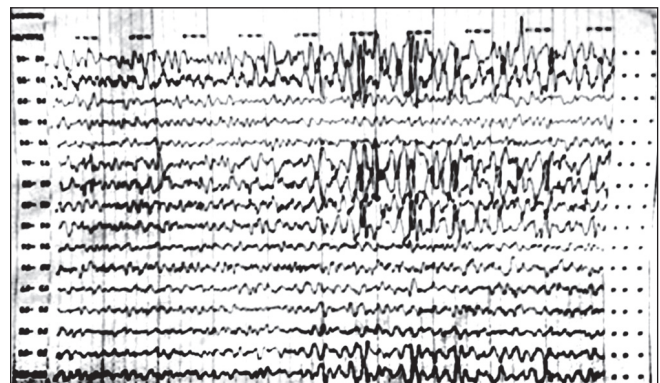


Figure 1: The EEG of Case no 5 showing paroxysmal spikes and waves discharge from left fronto-temporal leads with generalization

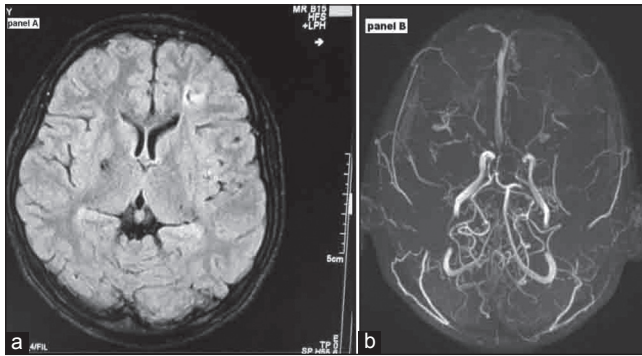


Figure 2: Contrast enhanced MRI brain (panel a) with MRA (panel b) of case no 6 showing narrowing of bilateral supraclinoid ICA, ACA and MCA

Discussion

Most children with moyamoya disease develop TIA or cerebral infarction. It generally causes cerebral ischemia in the territory of the ICA, particularly in the frontal lobe. Therefore, focal neurologic deficits like hemiparesis, dysarthria and aphasia are common manifestations.^[4] Case no 1, 2 and 6 represent the common ischemic manifestation of moyamoya disease in children. In Hokkaido Japan, 78.4% of the patients below 10 years of age had ischemic symptoms.^[5] In the United States, most children presented with ischemic symptoms.^[6] Moyamoya disease is an easily overlooked condition because of its low incidence, but it is an important cause of stroke in children. The patients who presented with ischemic symptoms in our case series were diagnosed having moyamoya disease only after they developed second clinical attack.

Moyamoya disease can present with visual disturbances.^[7] We had one patient presenting with amaurosis fugax with no abnormal findings, except MRI Brain with MRA suggestive of moyamoya disease. Such ocular symptoms with few abnormal ocular findings have been reported as manifestations of moyamoya disease.^[8]

Involuntary movements are important clinical presentations in moyamoya disease, particularly in pediatric patients.^[4] We had one case with prior history suggestive of ischemic stroke and presenting to us with hemichorea. Hemichorea as a manifestation of moyamoya has been reported from Japan and ischemia of the striatum has been suggested as a probable cause. All these reports included adults.^[9-11] We had a child presenting with hemichorea and notably her MRI brain with MRA had stenosis of bilateral PCAs in addition to ICA, ACA and MCA. Her symptoms responded to haloperidol and sodium valproate.

We had one child with focal seizures and on investigation, she was found to have abnormal EEG and findings suggestive of moyamoya disease on MRI brain with MRA. Although epilepsy is mentioned as an important manifestation of moyamoya disease, there are very few published reports.^[2,4] Very recently, a case of limb shaking TIAs and focal Jacksonian seizures in a young female was published.^[12]

Stenosis or occlusion of the PCA has been reported in 30.3% of patients with moyamoya disease. The hemisphere ipsilateral to PCA stenosis had higher incidence of ischemic symptoms, cerebral infarction, and impaired cerebral hemodynamics. The TIA (hemianopsia) or cerebral infarction in the occipital lobe was noted in some patients during follow-up, but progression of PCA stenosis was not noted in pediatric patients.^[13] However, a recent study showed progressive stenosis in 18% of the evaluated PCAs.^[14] We had two patients with PCA stenosis (one bilateral and one unilateral) and one of the hemichorea patient.

Because of its progressive nature, it is imperative to diagnose moyamoya disease early and offer surgical treatment.^[3] Owing to poor socioeconomic condition of our patients, they could not afford surgical revascularization and follow-up MRI.

Conclusion

Moyamoya disease may manifest as stroke in children. Apart from stroke, it may have other various clinical presentations. Owing to its low incidence in India, it is easily overlooked and may not be considered as a possible diagnosis for the clinical presentation. With the advent of advanced diagnostic facilities, more cases of moyamoya disease with different presentation may be identified.

References

1. Seol HJ, Wang KC, Kim SK, Hwang YS, Kim KJ, Cho BK. Headache in pediatric moyamoya disease: Review of 204 consecutive cases. *J Neurosurg* 2005;103:439-42.
2. Kikuta K, Takagi Y, Arakawa Y, Miyamoto S, Hashimoto N. Absence epilepsy associated with moyamoya disease. Case report. *J Neurosurg* 2006;104:265-8.
3. Han C, Chen J, Gao S, Duan L. Moyamoya disease in Asia. *Neurology Asia* 2012;17:175-81.
4. Kuroda S, Houkin K. Moyamoya disease: Current concepts and future perspectives. *Lancet Neurol* 2008;7:1056-66.
5. Baba T, Houkin K, Kuroda S. Novel epidemiological features of moyamoya disease. *J Neurol Neurosurg Psychiatry* 2008;79:900-4.
6. Scott RM, Smith JL, Robertson RL, Madsen JR, Soriano SG, Rockoff MA. Long-term outcome in children with moyamoya syndrome after cranial revascularization by pialsynangiosis. *J Neurosurg* 2004;100:142-9.
7. Miyamoto S, Kikuchi H, Karasawa J, Nagata I, Ihara I, Yamagata S. Study of the posterior circulation in moyamoya disease. Part 2: Visual disturbances and surgical treatment. *J Neurosurg* 1986;65:454-60.
8. Noda S, Hayasaka S, Setogawa T, Matsumoto S. Ocular symptoms of moyamoya disease. *Am J Ophthalmol* 1987;103:812-6.
9. Hama A, Furune S, Nomura K, Takada Y, Matsushima Y. A case of unilateral moyamoya disease presenting with hemichorea. *No To Hattatsu* 2001;33:166-71.
10. Kinboshi M, Inoue M, Kojima Y, Nakagawa T, Kanda M, Shibasaki H. Elderly case of moyamoya disease presenting with hemichorea. *Rinsho Shinkeigaku* 2012;52:25-9.
11. Miura T, Kobayashi M, Sonoo M, Isii K, Shimizu T. An adult case of Moyamoya disease presenting with transient hemichorea. *Rinsho Shinkeigaku* 2002;42:45-7.
12. Kraemer M, Diehl RR, Diesner F, Berlit P, Khan N. Differential diagnosis between cerebral ischemia, focal seizures and limb shaking TIAs in moyamoya disease. *Br J Neurosurg* 2012;26:896-8.

13. Kuroda S, Ishikawa T, Houkin K, Iwasaki Y. Clinical significance of posterior cerebral artery stenosis/occlusion in moyamoyadisease. *No Shinkei Geka* 2002;30:1295-300.
14. Lee JY, Kim SK, Cheon JE, Choi JW, Phi JH, Kim IO, *et al.* Posterior cerebral artery involvement in moyamoya disease: Initial infarction and angle between PCA and basilar artery. *Childs Nerv Syst* 2013;29:2263-9.

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