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Moyamoya Disease Masquerading as Hemiplegic Migraine in a Child: A Case Report from India

Author(s)

Kunal Kumar, Deepak Kumar, S. B. Mathur

Affiliation(s)

University of Delhi, Maulana Azad Medical College, Lok Nayak & Associated Hospitals, Department of Pediatrics, New Delhi, India

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Abstract

Moyamoya disease (MMD) is a chronic progressive steno-occlusive disease of the intracranial arterial vessels and their branches characterized by puff smoke appearance on angiography. Multifocal neurological insults and deficits are commonly observed in children with MMD. A 7-year-old boy presented with multiple episodes of transient hemiparesis preceded by headache, vomiting, and visual auras was initially labeled as sporadic hemiplegic migraine according to the International Headache Society criteria. However, the rare association of hemiplegic migraine with MMD compelled us to perform magnetic resonance imaging and digital subtraction angiography, which revealed MMD as the underlying cause for the alternating hemiplegia and headaches in the child.

Keywords: Moyamoya disease, alternating hemiplegia, hemiplegic migraine

Introduction

Moyamoya disease (MMD) is an infrequent disorder affecting the blood vessels in the brain and is characterized by narrowing of the distal internal carotid arteries (ICAs) and their branches. This narrowing progressively occludes the arteries in the circle of Willis, leading to the development of distinctive collateral vessels, which appear like a puff of smoke on angiography.[1] The average annual incidence of this condition is 0.54 per 100,000 populations and is prevalent among pediatric patients in Eastern Asia.[2]

Headache associated with MMD is a well-known presentation that persists even after revascularization surgery.[3] Although very few, there are reports stating that differential diagnosis of MMD should always be made in patients presenting with migraine-like headaches and alternating neuro deficits.[4-6] Here, we present an unusual case of MMD where the primary manifestation was alternating hemiplegic migraine.

Case Reports

A 7-year-old boy presented to a pediatric emergency department with the complaint of sudden onset weakness in the right side of the body and deviation of the mouth toward the left side. The event was preceded by auras consisting of headache, vomiting, bright sparkling lights in front of his

Correspondence: Deepak Kumar, University of Delhi, Maulana Azad Medical College, Lok Nayak & Associated Hospitals, Department of Pediatrics, New Delhi, India
E-mail: deepakk70@gmail.com **ORCID:** 0000-0002-3380-7009

eyes, and a sudden urge to urinate. The headache was throbbing and pulsatile in nature, of variable intensity, and resolved after a few hours, but focal neurological deficits continued to persist. On reviewing the history, there were three such episodes of left-sided body weakness preceded by aura in the past 9 months, which resolved within 6 h without any residual focal neurological deficits. Examination revealed: Glasgow coma score, 15/15; heart rate, 94; respiratory rate, 26; pulse, normovolemic and regular; blood pressure, 104 systolic and 78 diastolic; CFT, less than 2 s; fundus, no abnormality; there were no cranial nerve deficits; and right-sided hemiparesis was present (power of 2/5 by Medical Research Council scale). The rest of the neurological and systemic examinations were unremarkable. Family history was negative for epilepsy, migraine, or any psychiatric disorder. Clinically, the child was labeled sporadic hemiplegic migraine as per the International Headache Society criteria.

Hematological parameters: hemoglobin, 11 g/dL; total leukocyte count, 8,800; platelets, 2.8 lac/cumm; ESR, 10 mm; other vasculitis markers (ANA, anti-ds-DNA, C-ANCA, P-ANCA) were unremarkable; biochemical parameters: serum sodium, 142 mmol/L; serum potassium, 3.8 mmol/L; serum dextrose and ionized calcium, 80 mg/dL and 0.99 mmol/L, respectively. Arterial blood gas and urine drug/toxin screening were normal. Both electrocardiography and electroencephalography were normal for age. However, magnetic resonance imaging (MRI) brain with angiography showed acute to subacute infarction in the left parieto-occipital region with non-vascularization of bilateral ICA distal to the origin of ophthalmic arteries with filling defect in bilateral anterior cerebral artery and left middle cerebral artery through multiple lenticulostriate and thalamostriate collaterals, features suggestive of MMD, stage 3 (Suzuki classification) (**Figure 1 and 2**). Other investigations were ordered to rule out organic causes of headache with hemiplegia, i.e., mitochondrial disorder-MELAS, transient ischemic attack (TIA), and intracranial arteriovenous malformations. Subsequently, over the next 24 h, right-sided hemiparesis improved with power improvement from 2/5 to 4/5, with complete resolution of right-sided facial nerve deficit. Consent from both parents was obtained before the inclusion of the child for reporting the case, in accordance with the institutional ethics guidelines.

Discussion

Hemiplegic migraine is an unusual form of migraine with aura where the headache is accompanied by one-sided weakness that switches sides during or between attacks as well as visual auras. It can either run in families (familial hemiplegic migraine) or occur sporadically in an individual (sporadic hemiplegic migraine).[7,8]

In our case, the clinical presentation met the diagnostic criteria for sporadic hemiplegic migraine, characterized by reversible motor weakness and visual-sensory auras, as defined by the international classification of headache disorders (ICHD-3).[9] To rule out other potential causes that could mimic migraine, neuroimaging was performed, revealing MMD as the underlying condition.

MMD is a progressive narrowing and eventual blockage of blood vessels in the brain. The symptoms vary depending on the age of onset, with headaches, seizures, TIAs, and ischemic strokes being more common in childhood, whereas subarachnoid and intracerebral hemorrhages occur more frequently in adulthood.[10] Although headache and aura as the initial symptoms of pediatric MMD are rare, only a few case reports have described such occurrences.

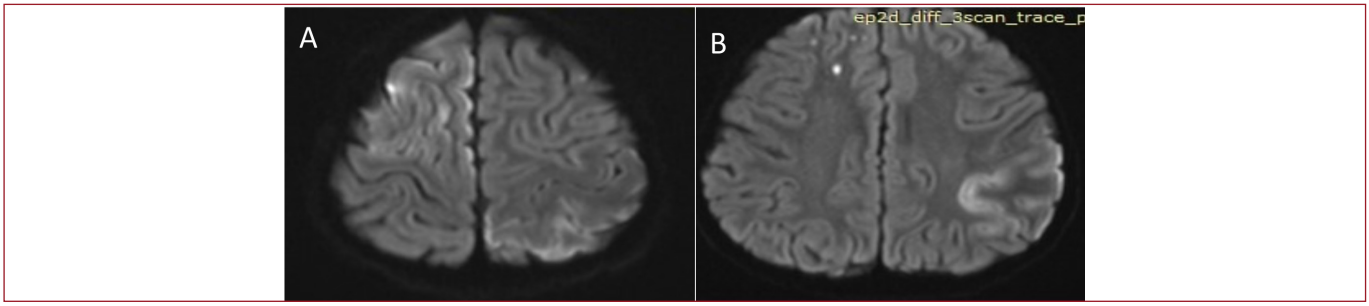

Figure 1. MRI Images (A,B) shows acute to subacute non-territorial infarct involving right frontal and left parieto-occipital region.
MRI; Magnetic resonance imaging

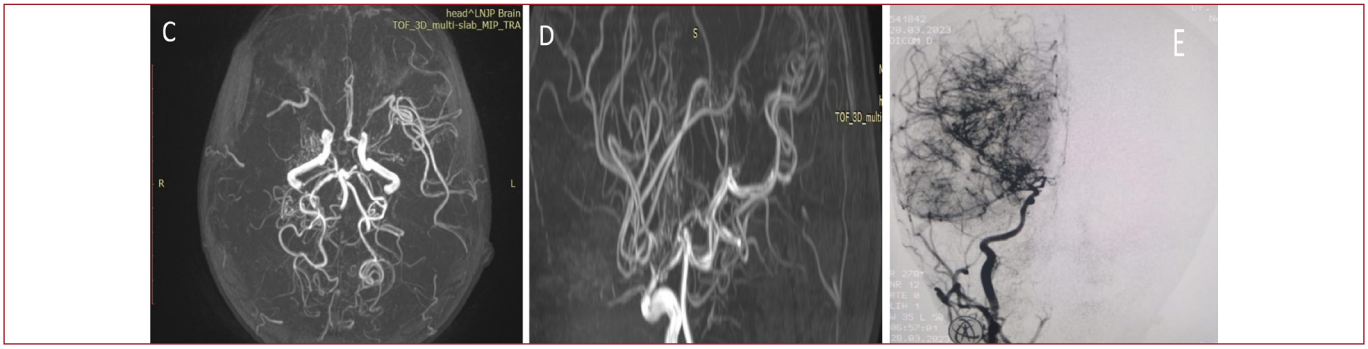

Figure 2. MRI brain (C,D) shows non-visualisation of B/L ICA distal to the origin of ophthalmic arteries with multiple collaterals of lenticulostriate and thalamostriate arteries (E) giving puff of smoke appearance.
MRI; Magnetic resonance imaging, ICA; Internal carotid artery

The first such occurrence was described by Bernstein[11] in 1993 in a 6-year-old girl. Cerebral artery nociceptor likely play a role in generating headaches in MMD, as studies have shown that stretch on these vessels can produce nausea and referred pain in regions associated with migraine attacks.[12] Neuroimaging techniques like computed tomography and MRI are useful in identifying organic or vascular causes in these patients, but cerebral digital subtraction angiography (DSA) remains the gold standard for diagnosing and monitoring MMD. DSA reveals classical angiographic changes, such as progressive stenosis and occlusion of the supraclinoidal part of the ICAs (usually bilateral) and the characteristic appearance of collateral vessels resembling a “puff of smoke”.

Conclusion

In conclusion, headaches associated with MMD can manifest as migraine with or without auras and may be the first and only presenting symptom. A high level of suspicion, consideration of various differential diagnoses, and early neurovascular imaging are crucial for diagnosis and prompt initiation of treatment.

Informed Consent: Consent from both parents was obtained before the inclusion of the child for reporting the case, in accordance with the institutional ethics guidelines.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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