

## Unusual association of diseases/symptoms

## Moyamoya disease presented as a case of hemiplegic migraine

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**Summary**

Moyamoya disease is a rare cerebral vasculopathy characterised by progressive narrowing of the major intracranial vessels and development of collateral vessels. Clinically, it presents with cerebral ischaemic or haemorrhagic events, with eventual severe morbidity and mortality. A 10-year-old girl presented with episodic vascular headache and transient left sided hemiparesis, which persisted for a few hours and was clinically labelled as sporadic hemiplegic migraine according to International Headache Society criteria. We investigated all possible causes of unusual headache. Cerebral digital subtraction angiogram was carried out and moyamoya disease was finally diagnosed.

**BACKGROUND**

Moyamoya disease is a chronic cerebral vasculopathy first described in 1957 by Takeuchi and Shimizu.<sup>1</sup> Progressive occlusion of the arteries in the Circle of Willis leads to development of the characteristic collateral vessels which are a network of pre-existing normal collaterals that have dilated in parallel with the occlusion of the Circle of Willis. Suzuki and Takaku observed that the collateral vessels give the appearance of a puff of smoke on arteriography and named the disease 'moyamoya' (puff of smoke in Japanese) in 1969.<sup>2</sup> Moyamoya involves the intracranial internal carotid arteries (ICAs) and their major branches: the anterior cerebral artery (ACA) or the middle cerebral artery (MCA). The posterior cerebral artery (PCA) and posterior communicating artery (PCoM A) can also be affected in advanced stages. The incidence of moyamoya disease among females is 1.8 times greater than among males and the highest incidence is found in Asia, especially Japan (0.35–2.0 cases per 100 000; prevalence 3.16).<sup>3</sup> Clinically, ischaemic events occur mainly in childhood, while haemorrhagic events occur in adulthood. Thus, moyamoya disease has a bimodal incidence with severe morbidity and mortality.

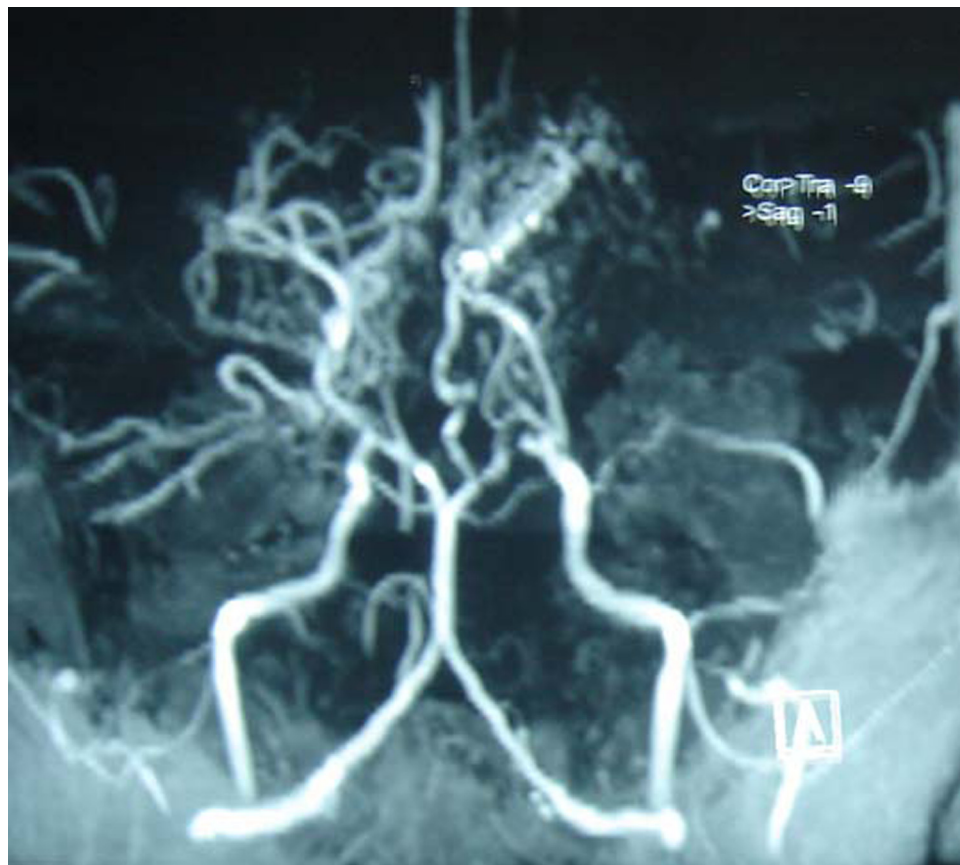
**CASE PRESENTATION**

A 10-year-old, right handed, Muslim girl from Moulvibazar was admitted to the Department of Neurology, Dhaka Medical College Hospital with the complaint of occasional left hemicranial headache for the last 5 years. The headache was throbbing, pulsatile in nature, of moderate intensity and associated with nausea, vomiting and photophobia. About 10–15 min before the headache, she saw bright sparkles of light in front of her eyes. The headache occurred every 1–2 months and persisted for few hours to 3 days. For the last 3 years, during some of the episodes she had also experienced transient weakness of the left side of her body, which started 15–30 min after the headache. This weakness usually persisted for 1–2 h and resolved spontaneously

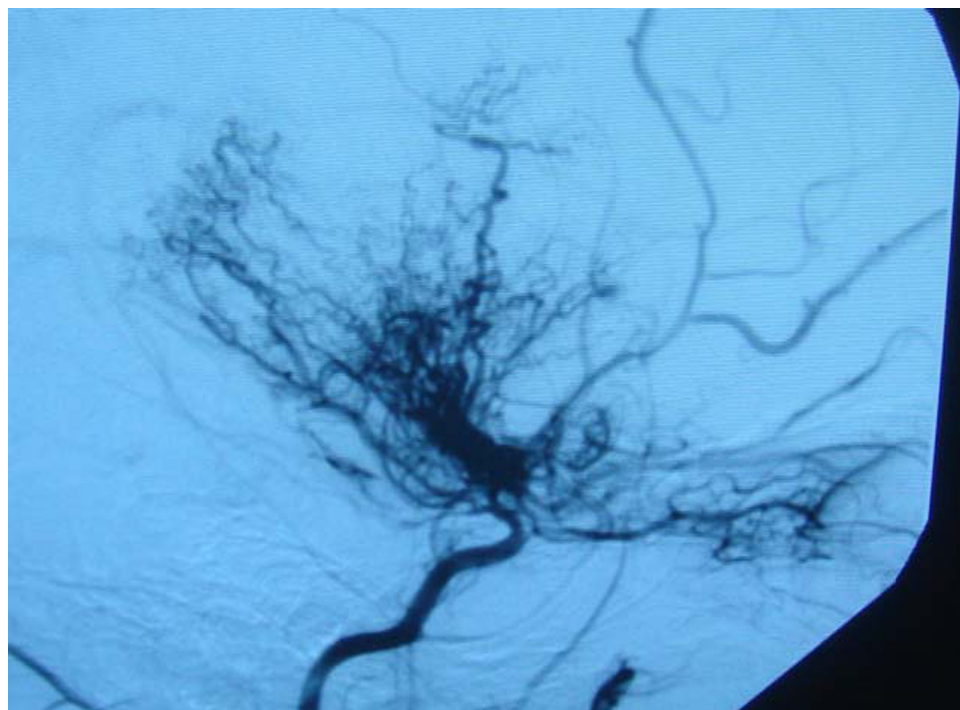
without any residual disability. She had no history of concurrent fever, trauma, convulsion, involuntary movement, change in speech, sensory impairment or loss of consciousness. She had no other significant past medical history. None of her family members have migraine-like headache. Physical examination was entirely normal and no neurological deficits were found. Clinically, she was diagnosed as having 'sporadic hemiplegic migraine' according to International Headache Society (IHS) criteria.

**INVESTIGATIONS**

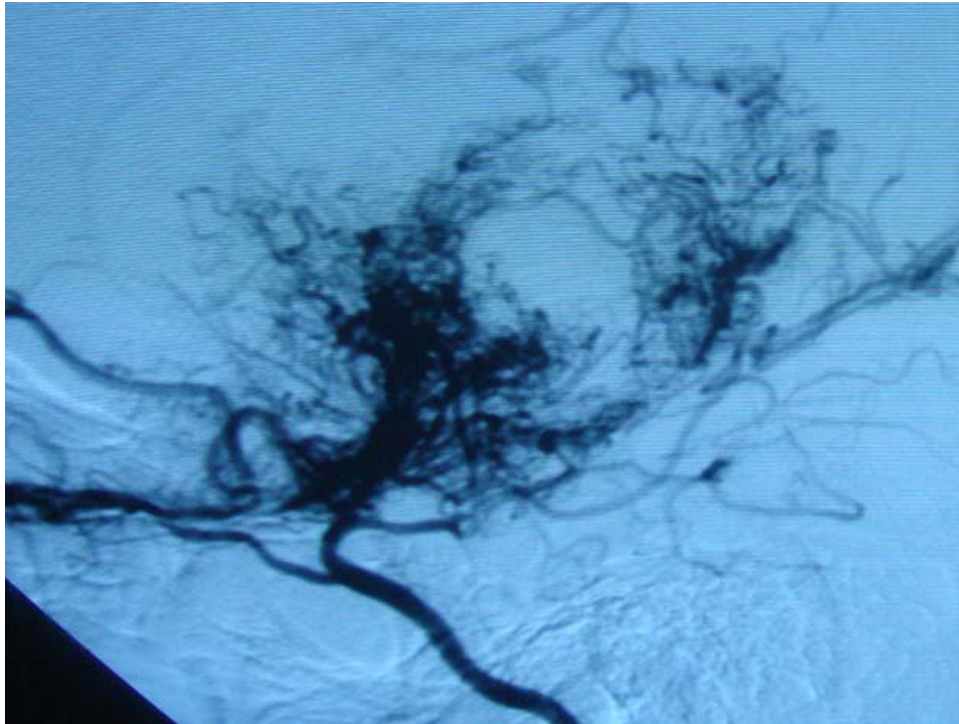
All her routine investigations and vasculitic markers (ESR, ANA, anti-ds-DNA, C-ANCA, P-ANCA) were unremarkable. However, MRI of the brain showed tiny flow voids in the basal ganglia region, predominantly in the left thalamus. Magnetic resonant angiogram (MRA) of the brain showed narrowing of the MCAs, ACAs, PCAs and their branches with poor visualisation of distal terminations (figure 1), suggesting moyamoya disease. These findings were confirmed by digital subtraction angiogram (DSA) of the brain which revealed discontinuation of the right ICA at the supraclenoid level after the PCoM A junction. Multiple dural and meningeal vessels supplied the right MCA and right ACA territory giving the typical smoky angiographic appearance. Right external carotid artery and left external carotid artery angiogram revealed multiple dural and meningeal twigs compensating the deprived area. Left ICA angiogram showed discontinuation of the left ICA just after the junction of the ophthalmic branch with the typical smoky angiographic appearance of moyamoya disease (figures 2 and 3). Left vertebral artery and right vertebral artery angiogram revealed normal findings with multiple choroidal and meningeal collaterals compensating the deprived ACA and MCA vascular territory. This sporadic hemiplegic migraine was finally correctly identified as a case of very rare moyamoya disease.



**Figure 1** Magnetic resonant angiogram of the brain reveals the middle cerebral artery, anterior cerebral artery and posterior cerebral artery and their branches are narrower with poorly visualised distal branches suggestive of Moyamoya disease.



**Figure 2** Cerebral digital subtraction RICA angiogram, lateral view, reveals discontinuation of the right internal carotid artery (arrow) at the supraclenoid level after the posterior communicating artery junction. Multiple dural and meningeal vessels supply the right middle cerebral artery and right anterior cerebral artery territory giving rise to the typical smoky angiographic appearance (dotted arrows).



**Figure 3** Cerebral digital subtraction LICA angiogram, lateral view, shows discontinuation of the left internal carotid artery (arrow) just after the ophthalmic branch junction with the typical smoky angiographic appearance (dotted arrows) of Moyamoya disease.

### DIFFERENTIAL DIAGNOSIS

Differential diagnosis of this headache with hemiplegia includes arteriovenous malformation with steal phenomenon, transient ischaemic attack (TIA) and mitochondrial disorder (such as MELAS).

### TREATMENT

The patient was treated symptomatically with non-steroidal anti-inflammatory drugs, propranolol and prochlorperazine with variable response. After final diagnosis we started nimodipine 30 mg three times daily and aspirin 75 mg daily.

### OUTCOME AND FOLLOW-UP

With treatment, the frequency and severity of headaches gradually subsided. The patient was reasonably symptom free for about 5 months but was then admitted again to our hospital with the same complaints in spite of continued treatment. This time she had ischaemic stroke with left sided hemiparesis. Subsequent MRI of the brain showed a small infarct in the right MCA territory.

### DISCUSSION

Diagnosis of hemiplegic migraine is based on a history of recurrent attacks of weakness associated with atypical headache, as in the present case. The clinical presentation of this case fulfils the diagnostic IHS criteria for sporadic hemiplegic migraine,<sup>4</sup> a form of complicated migraine.<sup>4</sup> In 1971, Verret and Steele described eight patients with hemiplegic migraine in childhood and one child with hemiplegic migraine at 3 months of age.<sup>5</sup> However, all cases

of sporadic hemiplegic migraine require neuroimaging and other advanced investigations to rule out other causes.<sup>4</sup> In our case we also sought to exclude other causes of migraine. MRI of the brain of our patient showed some signal change, which was later confirmed by MRA and cerebral DSA as a very rare case of moyamoya disease presenting as hemiplegic migraine.

In Japan, moyamoya disease is also called 'spontaneous occlusion of the circle of Willis'. In endemic areas, approximately 10% of local populations are genetically predisposed to the disease.<sup>6</sup> Linkage was found with loci on chromosomes 3p24.2-26 and 17q25 and on chromosome 6.<sup>6</sup> Clinical manifestation depends on age; headache, seizure, TIAs and ischaemic strokes are more common in childhood, whereas subarachnoid and intracerebral haemorrhage occur more frequently in adulthood.<sup>7</sup> There are two peaks in the age of presentation: below 10 and between 30 and 40 years of age, with the highest peak in the first decade and a lower peak in adulthood.<sup>8</sup> Only a few previous reports indicate that headache is a common symptoms in paediatric patients with moyamoya disease.<sup>7</sup> In 1993, Bernstein described an association between hemiplegic migraine and moyamoya disease in a 6-year-old girl.<sup>9</sup>

Imaging such as CT and MRI is useful to differentiate between ischaemia and haemorrhagic stroke. MRA is a non-invasive investigation to detect vascular abnormality,<sup>7 10</sup> but the gold standard for diagnosis and follow-up is cerebral DSA.<sup>7</sup> With DSA classical angiographic changes can be seen, such as progressive stenosis leading to occlusion of the supraclinoidal ICAs (usually bilateral) also involving the proximal MCA or ACA and numerous dilated perforating vessels in the base of the brain giving rise to the typical

'moyamoya' (puff of smoke) appearance.<sup>7 11</sup> Development of parenchymal, leptomeningeal or transdural collateral vessels can also be seen with additional collateral supply from branches of the external carotid artery and the ophthalmic system, all supplying the ischaemic brain.<sup>11</sup>

Early diagnosis and treatment are important during childhood as the disease can cause progressive disability, including deterioration in cognitive ability,<sup>12</sup> because viable cerebral tissue is necessary for reperfusion. The efficacy of the medical and surgical treatment of moyamoya disease is still controversial. In adults, treatment is aimed at improving cerebral ischaemia and preventing repeated intracerebral haemorrhage by reducing the overload on moyamoya collaterals.<sup>12</sup> In children the first line of treatment is pharmacological with anti-platelet therapy or calcium channel blockers, which can improve symptoms but do not halt disease progression.<sup>7 12 13</sup> Direct superficial temporal artery to MCA bypass is considered the treatment of choice, although its efficacy, particularly for haemorrhagic disease remains, uncertain. Multiple burr holes have been used in frontal and parietal lobes with good neovascularisation achieved. However, the most favoured approaches currently are the indirect procedures EDAS (encephalo-duroarterio-synangiosis) and EMS (encephalo-myo-synangiosis). It is very important to treat this condition promptly as once a major ischaemic or bleeding event takes place, even with treatment, the patient may be left with permanent loss of function.<sup>14</sup>

## Learning points

- ▶ All vascular headache with uncommon age and atypical presentations should be evaluated properly.
- ▶ Moyamoya disease is a rare central nervous system vascular anomaly more common in Asian populations.
- ▶ Hemiplegic migraine is a very uncommon presentation of moyamoya disease and we have found only a single previously reported case.
- ▶ Early diagnosis of the disease is important so that advanced management can be instituted.

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**Competing interests** None.

**Patient consent** Obtained.

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