Moyamoya disease; suspecting on conventional MRI brain without angiography

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Ann Indian Acad Neurol 2015;18:353-354

Clinical Description

A 10 year old, premorbidly normal girl presented with history of weakness of right half of the body, right focal seizures, and absent speech for 1 week. There was no associated history suggestive of encephalopathy, cranial nerve deficit, impairment of sensations or coordination, or raised intracranial tension. There was no prior history suggestive of transient ischemic attacks. Family history was unremarkable.

Salient features on examination were right hemiparesis with complete aphasia. There was no evidence of any neurocutaneous stigmata or dysmorphism. There was no other abnormality on general physical or systemic examination.

Her magnetic resonance imaging (MRI) brain showed left middle cerebral artery territory (MCA) infarct with attenuation of left internal carotid artery (ICA) flow void and presence of multiple flow voids around left ICA, suggesting presence of multiple collaterals hinting to diagnosis of Moyamoya disease. Magnetic resonance (MR) angiography done subsequently was confirmatory, revealing stenosis of supraclinoid left ICA with typical "puff of smoke" appearance of collaterals [Figures 1a and b].

Discussion

Moyamoya disease is a cerebrovascular condition characterized by progressive stenosis of distal ICA and/or MCA and anterior cerebral artery with compensatory formation of a network of perforating blood vessels providing collateral circulation. Moyamoya, in Japanese, means "hazy." The disease derives its peculiar name from the angiographic appearance of cerebral vessels in the disease that resembles a "puff of smoke." The process of narrowing of cerebral vessels seems to be a reaction of brain blood vessels to a wide variety of external stimuli, injuries, or genetic defects. Conditions such as sickle cell anemia, neurofibromatosis-1, Down's syndrome, congenital heart defects, antiphospholipid syndrome, renal artery stenosis, and thyroiditis have been found to be associated with Moyamoya disease.[1-3] Stenosis of ICA is visualized as attenuated flow void on MRI brain and the frequent collaterals appear as multiple flow voids around the ICA flow void. The same can be visualized as stenosis of ICA and " puff of smoke" like multiple collateral vessels on cerebral angiography.[4,5]

Digital subtraction angiography (DSA) is an invasive procedure compared to routine MRI brain. The emphasis here is on the fact that even in a child with unilateral focal neurological deficits, a meticulous evaluation of a non-invasive conventional MRI brain can points toward the diagnosis of Moyamoya disease. Early diagnosis and appropriate intervention is of paramount importance to prevent devastating long-term sequelae.

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	DOI: 10.4103/0972-2327.160080

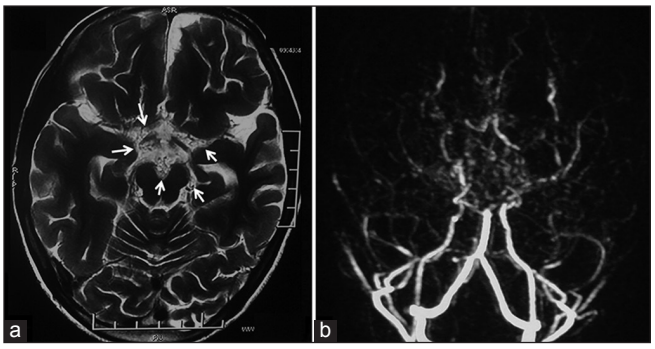

Figure 1: MRI brain showing left middle cerebral artery (MCA) territory infarct with multiple small tortuous flow voids in suprasellar and perimesencephalic cistern (arrows in Figure 1a) and loss of internal carotid artery and MCA flow voids hinting towards an underlying Moyamoya disease. Magnetic resonance angiography is confirmatory (Figure 1b)

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How to cite this article: Israni A, Chakrabarty B, Gulati S, Kumar A. Moyamoya disease; suspecting on conventional MRI brain without angiography. Ann Indian Acad Neurol 2015;18:353-4.
Received: 19-10-14, **Revised:** 20-11-14, **Accepted:** 04-12-14

Source of Support: Nil, **Conflicts of Interest**: None declared