Moyamoya disease: A case report with typical magnetic resonance imaging findings

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ABSTRACT

Moyamoya disease is an idiopathic non-progressive, non-atherosclerotic, and non-inflammatory vasculo-occlusive disease commonly affecting the supraclinoid internal carotid artery with resultant secondary changes which may include infarcts, ischemia, and multiple collaterals within the brain. The term Moyamoya disease is usually reserved for the idiopathic and rarely familial condition where etiology is not known, whereas the term Moyamoya syndrome is reserved for those conditions where etiology is known. Here, we present the case report of a Moyamoya disease and magnetic resonance imaging (MRI) findings with the review of the MRI features.

Key words: *Moyamoya disease, Ischemia, Angiography*

Moyamoya disease is an idiopathic non-progressive, non-atherosclerotic, and non-inflammatory vasculo-occlusive disease commonly affecting the supraclinoid internal carotid artery (ICA) with resultant secondary changes which may include infarcts, ischemia, and multiple collaterals within the brain [1]. Moyamoya disease derives its name from the Japanese which means the "puff of smoke." It leads to characteristic intracranial vascular changes. Moyamoya is a disease of children and young people with a female preponderance of 1:2 (male:female ratio) and a bimodal age distribution of 10 years and 30–40 years of age. The term "Moyamoya disease" should not be confused with Moyamoya syndrome where there are numerous list of causes including blood dyscrasias including sickle cell disease, polycythemia vera, infections, connective tissue disorders, phakomatosis, atherosclerosis, Marfan's syndrome, and Ehler-Danlos syndrome [1].

We report a case of 5-year-old female child who presented with a history of fall and weakness of the left upper limb and diagnosed as Moyamoya disease with typical imaging findings.

CASE REPORT

A 5-year-old female child presented to our emergency department with a history of headache, fall, and weakness of the left upper limb for 2 days duration. She had similar history 6 months back for which she was treated symptomatically by a local doctor. The headache was moderate in intensity with no diurnal variation and this was followed by dizziness and speech abnormalities. Her sensorium was normal with no history of fever, seizures, head injury, or sinusitis or visual disturbances. No neurocutaneous markers including skin pigmentations and angiomas were observed. Her general motor milestones were normal with normal development and no similar history was noted within the family.

On examination, her pulse rate was 80/min, BP 130/80 mmHg, temperature 98.5 Fahrenheit, and respiratory rate of 20/min. Her gait was hemiplegic with reduced power (2/5) in the left upper and lower limb, and plantar reflex was extensor on the left side with exaggerated reflexes. Cerebral spinal fluid analysis and peripheral smear were normal with normal hemogram and biochemical studies including renal and liver function tests.

On magnetic resonance imaging (MRI), ischemic changes were noted in the bilateral frontoparietal white matter. No evidence of acute infarcts (restrictions on diffusion-weighted imaging [DWI] and reduced apparent diffusion coefficient [ADC] values), and hemorrhage (blooming on gradient images) were observed (Fig. 1). On T2W images, multiple tiny dots/hypointensities were noted in the basal cisterna and Sylvian fissures representing dilated perforator vessels collaterals (Fig. 2). On MR angiogram, typical "puff of smoke" appearance was noted, with severe stenosis and occlusion of the bilateral supraclinoid ICA (Figs. 3 and 4).

A final diagnosis of Moyamoya was considered and the patient was treated symptomatically with aspirin 5 mg/kg for which she showed a slight improvement. Following symptomatic treatment, a superficial temporal artery-middle meningeal artery anastomosis was considered, following which, the patient showed a significant progressive improvement.

DISCUSSION

The vasculo-occlusive disease, Moyamoya affects the supraclinoid ICA and has unknown etiology. However, in a study done by

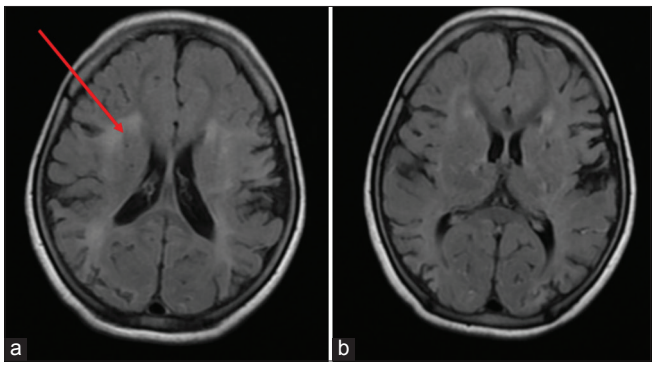

Figure 1: (a) Axial fluid attenuation inversion recovery (FLAIR) image showing ischemic changes in bilateral frontal (arrow) and parietal white matter; (b) Axial FLAIR image showing gliosis in the left parietooccipital lobe and lacunar infarcts in the left capsuloganglionic region

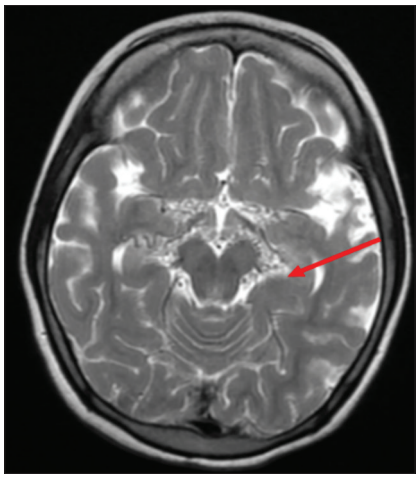

Figure 2: Axial T2W magnetic resonance image showing multiple flow voids in the basal cisterna (arrow) representing dilated collateral vessels

Bang *et al.*, they reported that genetic and environmental factors play a role in the development of the disease. These factors cause changes in the cytokines related to angiogenesis like caveolin which is a plasma membrane protein [2].

Clinically, the patients presented with ischemic symptoms and transient ischemic attacks including limb weakness, frequent falls, slurring of speech, difficulty in swallowing, and headaches. On MRI, multiple infarcts with restrictions on DWI and reduced ADC may be seen. Ischemic changes, gliosis, and Wallerian degeneration may be seen. On T2W images, multiple tiny flow voids representing the collaterals of the perforator vessels may be seen. On MRA, a classical puff of smoke due to collaterals from perforator vessels of multiple correspondence analysis and principal component analysis may be seen. There may be unilateral or bilateral occlusion of the distal supraclinoid ICA [3]. On angiogram, the same findings are seen.

Grading of the Moyamoya disease on the basis of severity as proposed by Suzuki and Takaku [4], it includes the following: Grade 1: Forking of ICA, Grade 2: Initiation of Moyamoya, Grade 3: Intensification of Moyamoya, Grade 4: Minimization of Moyamoya, Grade 5: Reduction of Moyamoya, and Grade 6: Disappearance of Moyamoya patterns are seen. In Grade 6, there is a disappearance of "puff of smoke pattern" with collateralization of intracranial circulation by external carotid artery (ECA).

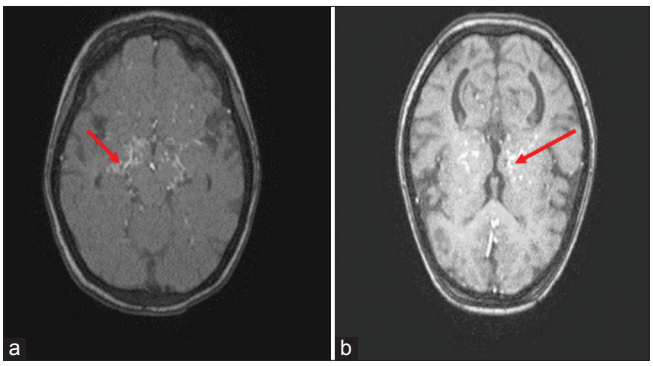

Figure 3: (a) Magnetic resonance (MR) angiogram source image showing multiple collaterals in basal cisterna (arrow), (b) T1W post-contrast MR image showing multiple collaterals in thalami (arrow)

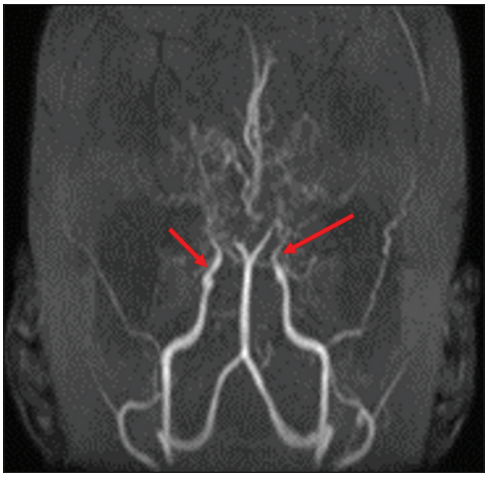

Figure 4: Magnetic resonance angiogram image showing severe stenosis of the bilateral supraclinoid internal carotid artery (arrows) with typical "puff of smoke" appearance

Treatment is usually surgical and pharmacological treatment includes control of hypertension and the antiplatelet agents may be given. Neurological rehabilitation and physiotherapy are supportive in case of infarcts. The various surgeries for Moyamoya disease include encephalomyosynangiosis (EMS), encephaloduroarteriosynangiosis, multiple burr holes for the development of collaterals from the scalp, and bypass procedures from the ECA and superficial temporal arteries [5-7]. Caldarelli *et al.* conducted a study on nine patients with Moyamoya disease and reported that if EMS surgery is done before significant cerebral damage occurs, it is very useful to stop the disease progression [8].

CONCLUSION

MRI with MR angiogram plays an important role in the diagnosis of Moyamoya disease, as well as, in the management and surgical planning of anastomosis.

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