

Case Report

MOYAMOYA disease-two case reports

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Introduction

Moyamoya disease is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels evidence by imaging, particularly cerebral angiography.¹ The disorder initially involves the intracranial portion of the internal carotid arteries and progresses to involve the middle, anterior and posterior cerebral arteries. Collateral vascular network developed due to compensatory dilatation of lenticulostriate and thalamostriate arteries. Angiographic study of these collateral vascular network give rise to characteristics appearance such as a puff of cigarette smoke. The Japanese term “MOYAMOYA” means “some thing hazy like a puff of cigarette smok.”²⁻⁴

Moyamoya disease was first described in Japan by Takeuchi and Shimizu in 1957. Though The disease is most common in Japan, many subsequent cases have been reported elsewhere Asia, Europe and North America. In 1988 the Japanese Ministry of Health and Welfare defined primary MMD as an idiopathic bilateral stenoses of arteries of the circle of Willis with collateral vascular networks, demonstrated on angiography.

The moyamoya syndrome (MMS) have same angiographic appearance as moya moya disease such as atherosclerosis, neurofibromatosis, postradiation therapy, sickle cell disease, systemic lupus erythromatosus and down syndrome (DS).¹

Case report-1

A three years old immunized girl of non consanguineous parents came to paediatric neurology of Bangabandhu Sheikh Mujib Medical University, Dhaka with left sided hemiparesis for 1 year. Mother stated that she developed seizure followed by left sided hemiplegia to extent that she unable to stand or walk. After this episode she gradually improved in standing & walking but weakness persist that made her walking difficult. Her perinatal and family history was normal. Milestone of development was age appropriate prior to this illness. There was no history of fever, unconsciousness, vomiting, or aphasia along with this illness and no history of radiation exposure or head trauma mentioned before this. On examination she had no facial dysmorphism, vitals were within normal limit, skin survey-normal, no chest murmur. Neurological findings revealed that she had hemiplegic gait, normal tone, power 4/5 on left side (both upper & lower limb) along with hyperreflexia,exaggrarated ankle clonus & babinski sign positive. Cranial nerve & sensory function, cerebeller function were intact.

Among the investigation, complete blood count, chest radiograph, ECG & EEG were normal. MRI of brain flare image showed bilateral chronic cerebral ischemic changes, more remarkable on right side. (Figure-1) MRA showed



Figure-1: MRI of brain (flare image) shows chronic cerebral ischemic changes, more remarkable on right side.

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gross narrowing of anterior cerebral, middle cerebral & posterior cerebral artery with extensive collaterals that appears as puff of smoke appearance suggestive of MOYAMOYA disease. (Figure-2 & 3) This patient was treated medically by nifedipine as vasodilator and ecospirin and patient become gradually ambulant. Then we planned to discharge the patient after neurosurgical consultation.

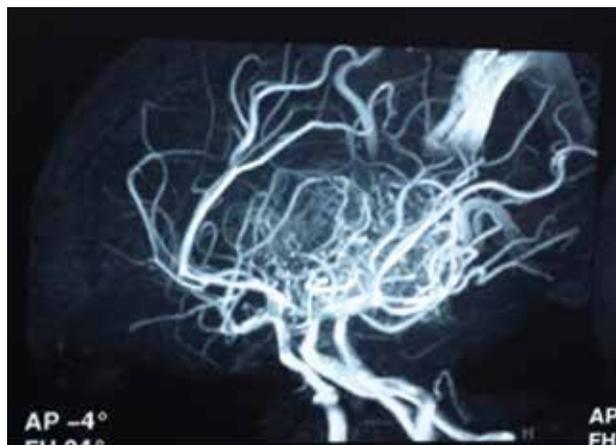


Figure-2: MR angiogram (right oblique view) shows characteristic Moyamoya vessels with typical “puff of smoke” appearance



Figure-3: MR angiogram (AP view) shows stenosis of bilateral internal carotid arteries

Case report- 2

A 5 years old immunized boy of consanguineous mating parents came from rural area of Bangladesh presented with left sided hemiparesis for last 5 days, was not associated with fever, unconsciousness or aphasia. He had

history of repeated attack of frontal, throbbing headache, associated with vomiting. He had an episode of chicken pox 1 month back. His perinatal history and milestone of development was normal. Examination findings reveals that he had no facial dysmorphism, vitals were within normal limit, multiple post varicella healed skin lesion found in whole body, no signs of meningeal irritation present there. Neurological findings –hypotonia, grade 3/5 power on left side (both upper & lower limb) with areflexia, bilateral plantar flexor, hemiplegic gait were present. Cranial nerve & sensory function, cerebellar functions were normal.

Among the biochemical reports, complete blood count, blood glucose, Lipid profile, serum lactate, Blood ammonia, renal function tests, liver function tests, CXR & ECG were within normal limit. CT scan of brain shows-small infarct in right parietal region. MRI of brain T2 and flair image multiple flow voids in the area of circle of willis and bilateral basal ganglia.(Figure-4&5) MR angiogram showed busy change in the both parietal region. (Figure-6&7) He was diagnosed as moyamoya disease. The patient was treated by methylprednisolone for 3 days followed by oral steroid along with aspirin. Patient was gradually improved & discharged with advice for regular follow up.

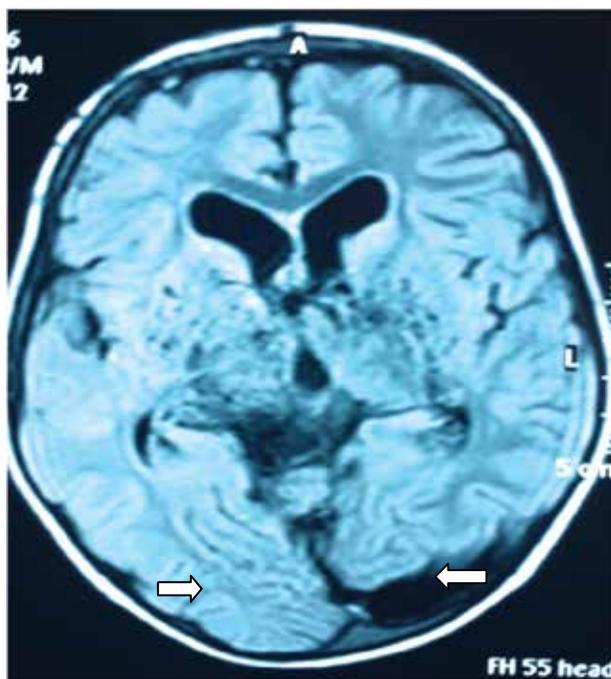


Figure-4: MRI of brain flair image showed multiple flow voids in basal ganglia region (arrow)

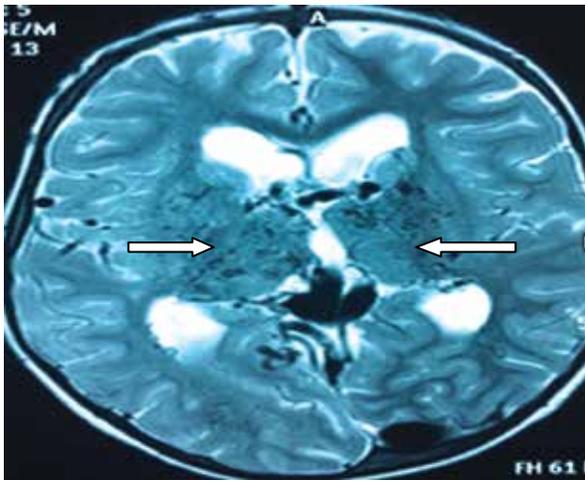


Figure-5: MRI of brain T2 image showed multiple flow voids in the bilateral basal ganglia(arrow)

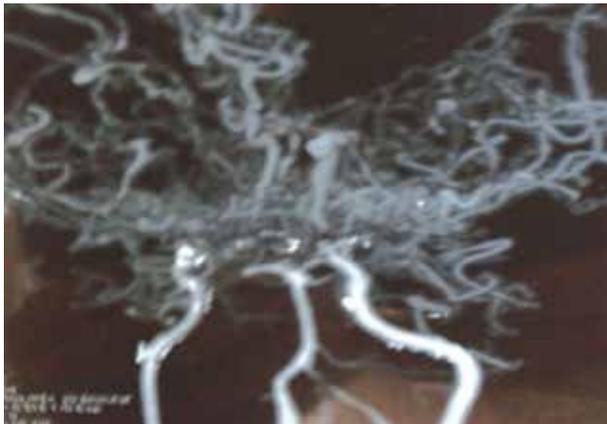


Figure-6: MR angiogram (A-P view) shows bilateral stenosis of internal carotid arteries with typical “puff of smoke” appearance



Figure-7: MR angiogram (right oblique view) shows characteristic Moyamoya vessels with typical “puff of smoke” appearance

Discussion

MoyaMoya disease is a rare idiopathic, slowly progressive vasculopathy characterized by irreversible stenosis of the arteries of Circle of Willis leading to cerebral hypoperfusion. The occlusive process stimulates the development of an extensive network of enlarged basal transcortical and transdural collateral vessels.

MoyaMoya disease was first reported in Japan in 1957 and has since been reported worldwide.⁴⁻⁷ The disease has a particularly high incidence in Eastern Asia, especially in Japan.⁶ The overall prevalence rate of moyamoya disease in Japan in 1995 was 3.16 per 100,000 with an incidence rate of 0.35 per 100,000.^{4,5} The male and female ratio is 1.8:1 and a family history of moyamoya is noted in 10% of cases.^{5,6}

The exact etiologic process of moyamoya disease is unknown, though myriad bacterial, environmental, genetic, and viral causes have been theorized.³

Family occurrence suggests that a genetic predisposition is present.^{4,5} Elevated levels of cellular retinoic acid-binding protein-I were found in the cerebrospinal fluid of patients with moyamoya disease.⁸ High levels of hepatocyte growth factor, a known inducer of angiogenesis, have been found in the carotid fork and cerebrospinal fluid of patients with moyamoya disease.⁹

Pathological changes in moyamoya patients include intimal thickening with fibrous tissue, abnormalities of internal lamina elastica, variable lipid deposition and virtual absence of inflammatory reaction in the blood vessel.^{10,11}

Clinically, the presentation of patients with moyamoya disease may include seizures, transient ischemic attacks, ischemic strokes, and hemorrhagic strokes.^{12,13,14} Visual deficits, speech disturbance, migraine like headache, intellectual deterioration, cranial nerve palsies, and disturbance of gait can also be evident.^{15,16} Our both patient presented with ischemic stroke and hemiplegia

Conventional angiogram or Digital subtraction angiogram (DSA) is the investigation of choice. MRA has a reliable diagnostic modality for MMD.⁷ CT and MRI are useful for detecting region of cerebral infraction. MRA defines the extent of disease and to some extent, the adequacy of collateral flow. It typically reveals the narrowing and occlusion of proximal cerebral vessels and extensive collateral flow through the perforating vessels demonstrating the classic ‘puff of smoke’ appearance.

T2W1 of MRI of our patient showed multiple slow voids at and around the base of the brain. Angiogram of both patient revealed stenosis of bilateral carotid systems as well as the typical 'puff of smoke' appearance of collateral vessels

Treatment in the acute phase is symptomatic with the goal of maintaining cerebro-vascular perfusion and function. Anticoagulant and antiplatelet agents have shown no remarkable benefit.¹⁷ The same lack of obvious efficacy has been described for corticosteroids in moyamoya disease.¹⁸ McLean et al, elucidated the use of verapamil hydrochloride to curtail the ischemic symptoms associated with moyamoya disease¹⁹, Surgical treatment modalities have been used to manage the hemorrhagic and ischemic consequences of moyamoya disease.^{4,10,20-22} Direct revascularization techniques, which are typically used in adults, include the superficial temporal artery to middle cerebral artery bypass or the middle meningeal artery to middle cerebral artery bypass are commonly applied in children.^{21,22}

Although Moyamoya is predominant in Japanese population but should not be overlooked in other population. The patient who will fulfill the clinical characteristics, MR angiogram should be done to diagnose Moyamoya disease.

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