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Case Report

Moyamoya disease: a case report

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ABSTRACT

Moyamoya disease is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels seen on cerebral angiography. The appearance of these small, multiple vessels at the base of the brain resembles "puff of smoke". The disease may develop in children and adults, but the clinical features differ. Children are likely to suffer with ischemic or haemorrhagic stroke. In the present report, authors describe a 7-year-old female with history of recurrent stroke apraxia and atrophy of left side with facial nerve palsy and was later on diagnosed with Moyamoya disease at HIMS Dehradun. Conclusion: Child with Moyamoya disease presents with recurrent episodes of sudden hemiplegia that might occur on alternate sides. The disease is progressive and dynamic in children. MRA clinches the diagnosis and neurosurgical intervention may be required as a part of definitive management.

Keywords: Moyamoya, Puff of smoke, Recurrent stroke

INTRODUCTION

Moyamoya disease is a rare, progressive cerebrovascular disorder caused by blocked arteries at the base of the brain in an area called the basal ganglia.[1] The name "Moyamoya" means "puff of smoke" in Japanese and describes the look of the tangle of tiny vessels formed to compensate for the blockage. The disease primarily affects children, but it can also occur in adults. In children, the first symptom of Moyamoya disease is often stroke, or recurrent transient ischemic attacks (TIA, commonly referred to as "mini-strokes"), frequently accompanied by muscular weakness or paralysis affecting one side of the body, or seizures. Magnetic Resonance Angiography (MRA) is the investigation of choice.[2-4]

CASE REPORT

Authors present the case of a 7-year-old female who visited the Emergency cell of Himalayan Institute of Medical Sciences, Dehradun with recurrent stroke and who was subsequently diagnosed as suffering from Moyamoya Disease on MRA.

A 7-year-old female presented in Paediatric emergency with left sided focal seizures associated with left sided facial muscle weakness causing facial asymmetry and difficulty in chewing from left side. She also had difficulty in doing daily light activities such as buttoning up, combing or wearing slippers. There was history of recurrent episodes of left sided hemiplegia, the first episode occurring at 3 years of age, with slowly progressive mental impairment.

On physical examination, general condition of the patient was stable, conscious, alert and oriented to time, place and person with apraxia. Anthropometry was appropriate for her age. On neurological examination the patient had UMN type of left facial nerve palsy and left hemiparesis with ataxia. Equal sensation was noted in bilateral upper

and lower extremities. The bulk and power decreased with facial atrophy of left side along with the atrophy of distal muscles on left side of both upper and lower limb. Cardiovascular and rest of the systemic examination seemed to be within normal limits.

Results of complete blood cell count showed normocytic anaemia with normal leukocytes and platelet counts along with coagulation profile being normal. Specific tests for hypercoagulability such as APLA, ANA antibodies, basal homocysteine levels were also assessed, the results for which were normal. An EEG was suggestive of bilateral parietal occipital slowing. An MRI brain was done which revealed a hyper intense signal in cortex in right temporal and parietal lobe with similar signal intensity seen on left side. In addition, there was no abnormal enhancement or mass or any acute infarct. Magnetic Resonance Angiogram of brain revealed no visualisation of ICA artery with multiple collateral formations, on the basis of which a diagnosis of Moyamoya disease was made (Figure 1). Patient was started on oral aspirin and neurosurgical opinion was sought. However, the patient was lost to follow up.

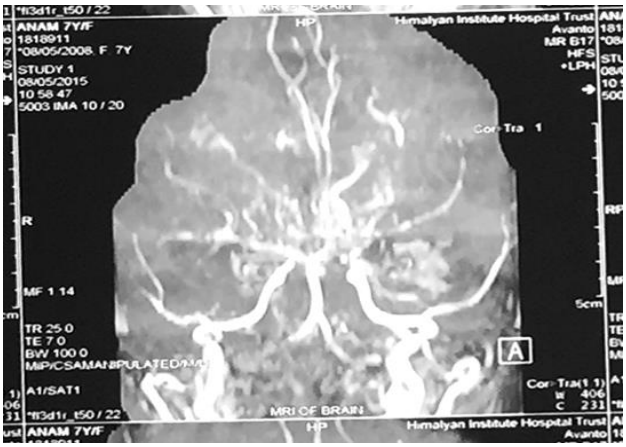

Figure 1: MRA of Brain.

DISCUSSION

Arteriopathy refers to disorders of the cerebral arteries and is a leading cause of childhood AIS. One such entity is Moyamoya disease (a Japanese word meaning a puff of smoke drifting in air). It is a progressive disorder associated with occlusion of the internal carotid artery at the terminal bifurcation together with abnormalities of anterior and middle cerebral arteries. These changes are bilateral. The presence of abnormal blood vessels at the base of the brain on MRA, give a hazy like puff of cigarette smoke drifting in air.[5] The process of narrowing of blood vessels is an inflammatory reaction of brain blood vessels to a variety of external stimuli, injuries or genetic defects. Moyamoya disease was first described in Japan by Takeuchi and Shimizu in 1963.[5,6] The disease manifests like a syndrome associated with other conditions such as neurofibromatosis type 1, Trisomy 21, Alagille syndrome, sickle cell anaemia, iron deficiency anaemia, coagulation disorders (including both hereditary and acquired), chromosomal microdeletions /micro duplications and post irradiation. Congenital malformations of the cranio cervical arteries including PHACES Syndrome or fibromuscular dysplasia may also lead to this abnormality.[7]

Moyamoya disease has been recognized as a cause of ischemic and haemorrhagic strokes.[7] The process of blockage is continuous despite medical treatment. The clinical features of moyamoya disease differ between children and adults. In contrast to adults, who typically present with subarachnoid or intra parenchymal haemorrhage, children with Moyamoya disease usually exhibit recurrent transient ischemic attacks (TIA) or infarction in the territory of the ICA, particularly in the frontal lobe. The signs and symptoms include mono paresis, hemi paresis, aphasia, and dysarthria. In addition to these - Headache, seizures, and involuntary movements, such as hemi chorea, are serious symptoms associated with paediatric moyamoya disease. Hyperventilation worsens the symptoms and is also the triggering factor.[6,7]

The confirmatory tool to diagnose the disease after MRI is Magnetic Resonance Angiography (MRA). It reveals narrowing and occlusion of proximal cerebral vessels with blood flowing through collaterals giving puff of smoke appearance.[8]

The progression of Moyamoya follows a typical course and can be classified into stages based on angiography findings.

Stage I: Narrowing of internal carotid arteries

Stage II: Development of moyamoya vessels at the base of the brain

Stage III: Intensification of moyamoya vessels and internal carotid artery narrowing (most cases diagnosed at this stage)

Stage IV: Minimization of moyamoya vessels and increased collateral vessels from the scalp

Stage V: Reduction of moyamoya vessels and significant internal carotid artery narrowing

Stage VI: Disappearance of moyamoya vessels, complete blockage of internal carotid arteries, and significant collateral vessels from the scalp.[8,10]

The management of Moyamoya is multifaceted. It includes antithrombotic strategies and hyper acute neuroprotective strategies essential to prevent the progressive ischemia. Antiplatelet drugs help to prevent future ischemic strokes as they inhibit platelet function and its use is considered empirical. Aspirin is effective if given within 48 hours of stroke or TIA at a dose of 50-

325 mg/day orally then 75-100 mg/day per orally. Neuro protective strategies include reduction of intracranial pressure by reducing cerebral blood flow and by overcoming the underlying triggering factors.[8] Long term prevention is revascularisation surgery. Surgical procedures are classified into three categories, direct bypassing including superficial temporal artery to MCA (STA-MCA) anastomosis, indirect bypassing including encephaloduroarteriosynangiosis (EDAS) and encephalo myosynangiosis (EMS), and combined bypassing. Indirect bypass surgery is a simpler technique and induces spontaneous angiogenesis within 3-4 months.[9,10]

CONCLUSION

A child with Moyamoya disease presents typically with recurrent episodes of sudden hemiplegia that might occur on alternate sides. The disease is progressive and dynamic in children. MRA clinches the diagnosis and neurosurgical intervention may be required as a part of definitive management.

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