

CASE REPORT

A SUCCESSFULLY TREATED CASE OF MOYA MOYA DISEASE

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HOW TO CITE THIS ARTICLE:

P. Yashodhara, N. Madhavi, K. Rami Reddy, B. Deeva Kumar, B. Elizabeth. "A Successfully Treated Case of Moya Moya Disease". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 04, January 12; Page: 702-706, DOI: 10.14260/jemds/2015/100

ABSTRACT: Moyamoya disease is a rare cerebrovascular disease characterized by progressive occlusive cerebral arteritis affecting the distal internal carotid arteries near the circle of Willis. A collateral circulation develops around the blocked vessels which appear as a "puff of smoke". Incidence is variable depending on the geographical location. The clinical features of Moyamoya disease are recurrent transient ischemic attacks (TIAs), strokes, sensorimotor paralysis, convulsions and/or migraine-like headaches. Magnetic resonance imaging (MRI) and MR Angiogram (MRA) should be performed for the diagnosis and follow-up of Moyamoya disease. Neurosurgical procedures like Encephalo myo synangiosis (EMS), multiple burr holes and some direct procedures are useful to reestablish new circulation after a few weeks. The long term outlook for patients with treated Moyamoya seems to be good. Once major stroke or bleeding take place, even with treatment, the patient may be left with permanent loss of function. So it is very important to treat this condition promptly. Herewith, we are reporting a 15 month old child with history of right focal adverse seizures, 3- 4 events a month followed by post ictal drowsiness. Imaging studies revealed a vascular insult with infarct in the left fronto parietal lobes as well as multiple infarcts in other regions of the brain. MR Angio Brain revealed Moya moya disease. Child was successfully managed by prompt referral to Neurosurgical Centre where he underwent EMS and he is under their follow up.

KEYWORDS: Moya moya disease, arteriopathy, puff of smoke, Encephalo myosynangiosis.

KEYMESSAGE: Moya moya Disease is a rare disease which is prevalent in Asian population. It is a rare but treatable disorder. Though the prognosis is poor, early diagnosis and prompt referral for surgery improves the outcome.

INTRODUCTION: Moyamoya is a rare cerebrovascular disease associated with approximately 6% of childhood strokes.^{1,2,3} Recognised in Japan in the 1960s, this is a progressive occlusive cerebral arteritis affecting the distal internal carotid arteries near the circle of Willis. Initially thought to be a disease occurring primarily in Asian countries, the number of patients diagnosed in Europe^{4,5} is increasing due to increased awareness of the condition. Moya moya disease in children usually presents with transient ischemic attack or recurrent stroke and prognosis is variable. Early diagnosis and management with recent surgical procedures like EMS improves outcome in children. We are reporting a one year old female child with right hemiparesis which was diagnosed as Moya moya disease and managed surgically so as to improve the long term outcome.

CASE REPORT: A One year old female child born out of second degree consanguineous parentage presented with history of seizures, 3-4 events per month followed by paucity of right sided movements. Child was apparently well up to 3 months of age, when her mother noticed right focal seizures & forceful sustained turning of head to right side. Repeated episodes occurred for every 3 months till 12 months of age. She was never evaluated and treated. Past history and birth history was

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not significant. Family history showed elder sibling having Generalized tonic clonic seizures secondary to Hypoxic ischemic encephalopathy. Growth and development, vitals and general physical examination of the child was normal. Central nervous system showed right sided hemiparesis and other systems were normal. Child was investigated for possibility of sickle cell anemia, neurofibromatosis and Moya moya disease. MRI Brain showed multiple foci of hyper intensities bilaterally. MRA showed stenotic lesions involving the supraclinoid and paraclinoid segments of both right and left internal carotid arteries extending into the M1 segment of middle cerebral arteries. Collateral supply to anterior cerebral arteries was present through the ethmoidal branches of ophthalmic arteries. Vertebro basilar circulation revealed the presence of multiple collaterals in the deep regions of cerebral hemispheres. In view of the above findings, definitive diagnosis of Moya moya disease was made. Child underwent Encephalo Myo Synangiosis (EMS) and now under the follow-up of the Neurosurgical team.

DISCUSSION: Moya moya disease is a chronic progressive non-atherosclerotic non-inflammatory non-amyloid occlusive intracranial vasculopathy of unknown cause. This disease was first reported by Takeuchi and Shimizu in 1957. The term Moyamoya disease was first used in 1969 which means “puff of smoke” used to describe the collateral circulation at the base of the skull. It is more common in the Asian population. Overall incidence is 1 per 1,00,000 and Male: Female ratio is 1: 1.65. Peak age of onset of disease is bimodal, with an early peak occurring in the first decade of life and a second peak in the fourth decade of life. Areas of several different chromosomes have been shown to contain genetic mutations which may cause Moya moya disease. These areas are MYMY1 on chromosome 3 (3p26-p24.2), MYMY2 on chromosome 17 (17q25.3), MYMY3 on chromosome 8 (8q23), MYMY4 on the X chromosome (Xq28), MYMY5 on chromosome 10 (10q23.3) and a further area on chromosome 6.⁶⁻¹² Moya moya disease can present either as transient ischaemic attacks (TIAs) (more common with children) or stroke (haemorrhagic - more typical in adults). Children usually present with headache, hemiparesis, seizures, disturbed consciousness, speech deficits (aphasia), sensory and cognitive impairments, involuntary movements and vision disturbances. Pathologically there is fibrocellular intimal thickening, smooth muscle cell proliferation and increased elastin accumulation resulting in the stenosis of suprasellar intracranial internal carotid arteries. The gold standard test for diagnosing Moya moya is radio-imaging with CT scanning or MRA showing stenosis or occlusion at the terminal portion of the internal carotid artery or the proximal portion of the anterior or middle cerebral arteries with abnormal vascular networks in the vicinity of the stenosed areas.

There is no role of antiplatelet drugs or anticoagulants in the management. Since Moyamoya tends to affect only the internal carotid artery and nearby sections of the adjacent anterior and middle cerebral arteries, other arteries, such as the external carotid artery or the superficial temporal artery can be directed to replace the blocked circulation. Currently the most favored surgeries are the in-direct procedures Encephalo duro arterio synangiosis (EDAS), EMS, and multiple burr holes and the direct procedure superficial artery – Middle cerebral artery (STA-MCA). The single greatest predictor of overall outcome for patients with Moyamoya is the neurologic status at time of treatment.^{13,14} Even if severe angiographic changes are present, if surgical revascularization is performed before disabling infarction, the prognosis tends to be excellent.

In our index case, the child was promptly referred for surgery to improve the chances of better long term outcome.

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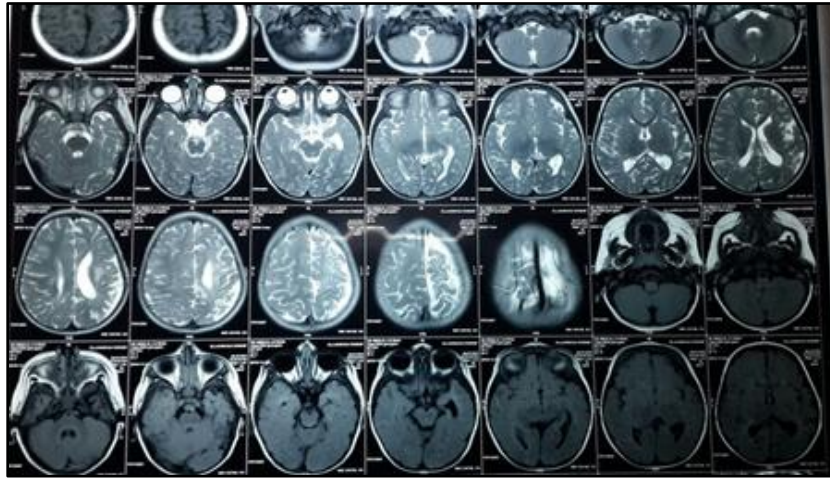


Fig. 1: MRI brain

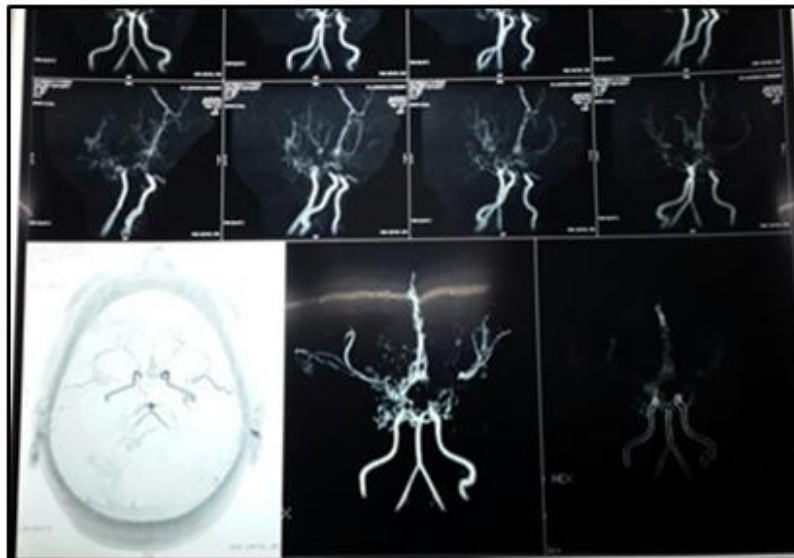


Fig. 2: Cerebral angiogram

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Date of Submission: 02/01/2015.
Date of Peer Review: 03/01/2015.
Date of Acceptance: 07/01/2015.
Date of Publishing: 10/01/2015.