Moya Moya Disease Masquerading as Transient Ischaemic Attacks

Mona Gajre*, Surbhi Rathi**, Ranjit Torgulkar***, Trimurti Nadkarni#, Mukesh Agrawal+

Abstract

Moya moya disease is a rare, idiopathic, irreversible arteriopathy of major blood vessels to the brain. In Japanese it means “puff of smoke” which refers to the collateral circulation and the disease can lead to severe functional impairment or even death.1,2 The neuroimaging of choice is magnetic resonance angiography (MRA) and it is better than other diagnostic modalities.3,4 The only known treatment is surgery the goal being to improve blood flow to hypoperfused cerebral regions. Surgical procedures such as direct bypass superior temporal artery to middle carotid artery anastomosis (STA-MCA) and indirect bypass like Encephaloduroarteriosynangiosis (EDAS) have shown excellent prognosis.

Introduction

Moya moya disease is a unique cerebrovascular disease with a high incidence in Japan and Asia. The disease was first described by Suzuki in 1963 and the surgical management began in the mid 1970’s. The Research Committee on spontaneous occlusion of the circle of Willis (Moya moya disease) of the Ministry of Health and Welfare, Japan, has studied the pathogenesis, epidemiology, clinical investigations and treatment of the disease since 1977.1 We present a case of moya moya disease with an unusual symptomatology of transient ischaemic attacks.

Case Report

An 8 year old boy was brought with a history of paroxysmal episodes of brief loss of power in right upper and lower limbs since 3 years. Initially the attacks were every six months but since last one year have progressed to every 3-4 months. The episodes were not associated with loss of consciousness or any neurological deficit and complete recovery present. Neuroinvestigations revealed a normal CT scan, EEG showing generalized cerebral dysrhythmia. The child was started on two anticonvulsants without symptomatic relief. On witnessing the episode in the wards it was diagnosed as transient ischaemic attacks (TIA) and on clinical suspicion a magnetic resonance angiography (MRA) was ordered. It revealed grade 3 moya moya disease with moderate to severe narrowing of the left intracranial portion of internal carotid artery (ICA), left middle carotid artery (MCA), segments of both anterior carotid artery (ACA) and posterior carotid artery (PCA), multiple collaterals in left thalamus and perimesencephalic cistern. Digital Subtraction Angiography (DSA) showed chronic progressive occlusive arteriopathy affecting (L) ICA, ACA and both PCA. An indirect bypass surgery Encephaloduroarteriosynangiosis (EDAS) anastomosis was done. Patient is symptom free post procedure since 3 months and is continued on low dose aspirin as a preventive therapy.

Discussion

The exact aetiology of Moya Moya disease is unknown. There is a female dominance with a male to female ratio of 1:1.7. An increased incidence is seen in children less
than 10 years of age and a second mild peak noticed from 30-40 years. The disease is of primary and secondary type. The former is inherited and the latter is found to be associated with conditions like Down’s syndrome, tuberous sclerosis, sickle cell disease, connective tissue disorders like Polyarteritis nodosa (PAN), Kawasaki’s disease and systemic lupus erythematosus (SLE), neurofibromatosis-1, Suprasellar tumours. Pathologically the disease consists of intimal wall thickening of the terminal portions of unilateral/bilateral internal carotid vessels. When unilateral the diagnosis is ‘probable moya moya’ whereas bilateral involvement is definite disease. Other brain vessels like the ACA, MCA and PCA may also show varying degrees of stenosis or occlusion.

The disease can be categorized into the following stages on the MRA (Suzuki’s Classification)

- **Stage 1** – Narrowing of the ICA bifurcation
- **Stage 2** – ACA, MCA, PCA dilated.
- **Stage 3** – Maximum basal moyamoya collaterals, small ACA/MCA
- **Stage 4** – Fewer collaterals, small PCA.
- **Stage 5** – Further reduction in collaterals, absent ACA, MCA, PCA
- **Stage 6** – Extensive pial collateral from external carotid branches.

Clinical presentation is of recurrent alternating hemiparesis, TIA’s, headache and aphasia. Children with this disease usually have strokes, whereas adults have ischaemic symptoms or intracranial haemorrhage. The natural history of untreated moyamoya disease is poor, with a 73% rate of major deficit or death more than 2 years after diagnosis in children and a similarly poor prognosis in adults. Newer modalities like MRI and MRA have made it possible to obtain a diagnosis without conventional angiography. Studies to assess cerebral perfusion and metabolism by positron emission tomography (PET) or single positron emission computed tomography (SPECT) are assuming great significance in understanding the state of illness and surgery. Management...
of moya moya disease can be either medical or surgical. The former includes antihypertensives, anticoagulants and antiplatelet drugs. Surgical therapies consists of either direct bypass surgery (STA-MCA anastomosis) or indirect bypass procedures such as Encephaloduroarteriosynangiosis (EDAS), EMS, encephalo-duro-arterio-myosynangiosis (EMAS) and omental transplantation. At present the surgically-treated groups have better results than the medically-treated groups.2,3

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References