

# Moya Moya Disease - A Rare Case of Cerebro Vascular Disease

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## Abstract:

*Moya Moya Disease* <sup>[1,3]</sup> is a chronic progressive non atherosclerotic non inflammatory non amyloid occlusive intracranial vasculopathy of unknown cause. Incidence is 1 per 1,00,000. In our case, 15 months baby presented with history of seizures, 3-4 events per month followed by paucity of right sided movements. CT Brain showed large areas of post ictal edema in the left fronto - parietal lobes and gliotic areas in the right parieto - occipital lobes. MR Angiogram with Right Carotid injection showed supraclinoid occlusion with collaterals and Vertebro Basillar injection showed multiple collaterals giving Puff of Smoke <sup>[2]</sup> appearance which is diagnostic of Moya Moya Disease. Child was treated surgically by Encephalo Myo Synangiosis <sup>[4]</sup> (EMS).

**Keywords:** Moya Moya Disease, MR Angiogram, Puff of Smoke, Encephalo Myo Synangiosis.

## I. INTRODUCTION

Moyamoya is a rare cerebrovascular disease. Recognised in Japan in the 1960s, this is a progressive occlusive cerebral arteritis affecting the distal internal carotid arteries near the circle of Willis. A four-year study in Japan from 2002-2006 found that the detection rate per year was 0.94 patients per 100,000 people. The prevalence was 10.5 patients per 100,000. There was a female to male ratio of 2.18:1. The incidence of asymptomatic patients was 17.8%.

There were two peaks of onset between 45 and 49 years and between 5 and 9 years. Initially thought to be a disease occurring primarily in Asian countries, the number of patients diagnosed in Europe is increasing due to increased awareness of the condition.

## II. CASE REPORT

A 15 month old child, second in birth order born out of consanguineous parentage was brought with history of right focal adverse seizures, 3- 4 events a month followed by post ictal drowsiness. 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. Never evaluated and never treated. At 13<sup>th</sup> months of age had 4 events of generalized tonic clonic seizures, for which she was evaluated and

diagnosed to have the present problem. There is also history of delayed milestones. No history of TB contact, Bronchial asthma, Trauma, recurrent respiratory infections, stroke, recent surgeries, blood transfusions, recent vaccination. Her elder brother aged 4 years is a patient of Generalized Tonic Clonic Seizures semiology due to hypoxic ischaemic encephalopathy secondary to birth asphyxia. Her younger brother aged 5 months was diagnosed case of severe myoclonic epilepsy of infancy probably Dravet Syndrome .

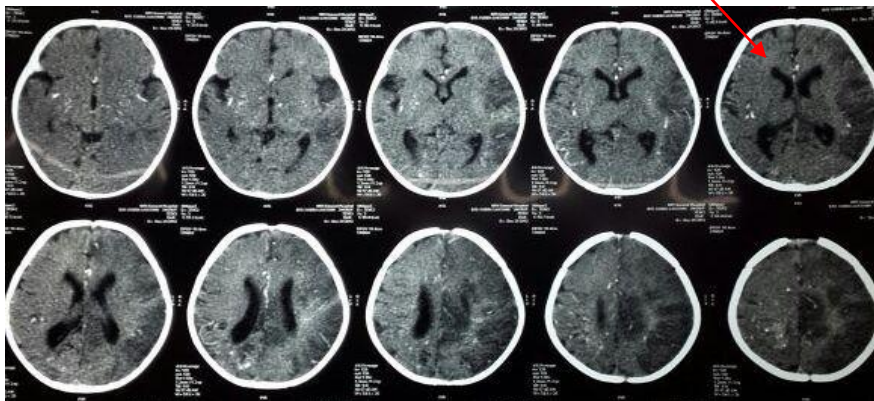
General Examination revealed: Child was conscious, alert. Temperature – Afebrile (98.4 F), Pulse rate – 110/ minute, Respiratory rate – 34 / minute, Blood pressure – 90/60 mm of Hg. In Nervous System Examination: All Cranial nerves are normal except for Mild right Upper Motor Neuron Facial palsy. Motor system: Bulk, Tone, Deep Tendon Reflexes were normal. Power: 3/5 in Right Upper Limb & 4/5 Right Lower Limb.

Complete Haemogram, Erythrocyte Sedimentation Rate (ESR), Protein C, Protein S, Antithrombin III, Factor – V levels, Electro Encephalogram (EEG) were normal. CT Brain showed large areas of post ictal edema in the left fronto - parietal lobes and gliotic areas in the right parieto - occipital lobes. (Fig- 1). MR Angiogram showing severe stenosis in the distal Internal Carotid Artery in Supraclinoid segment. (Fig -2)

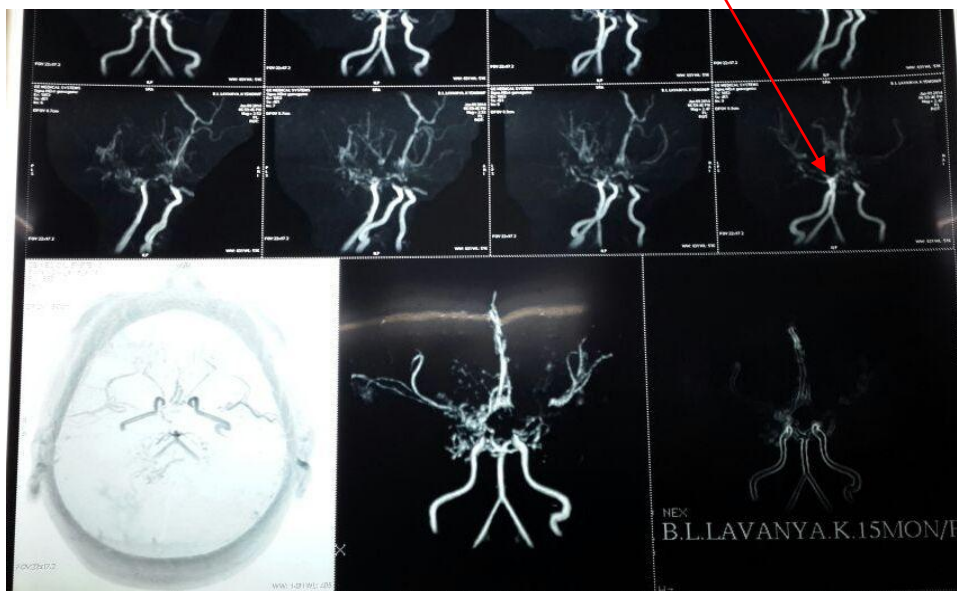
Cerebral Angiogram showed the long segment stenotic lesions involving the supraclinoid and paraclinoid segments of both right and left internal carotid arteries extending into the M1 segment of right and left middle cerebral arteries. Collateral supply to the right and left anterior cerebral arteries was present through the ethmoidal branches of right and left ophthalmic arteries. (Fig -3) Vertebro basilar circulation revealed the presence of multiple collaterals in the deep regions of cerebral hemispheres through the posterior choroidal and thalamogeniculate collaterals. (Fig -4). Cerebral Angiogram gave Puff of Smoke appearance which is due to collateral formation and diagnostic of Moya Moya Disease. (Fig- 5).

Child was treated surgically by Encephalo Myo Synangiosis (EMS).

CT Brain showing post ictal edema in the left fronto - parietal lobes and gliotic areas in the right parieto – occipital lobes



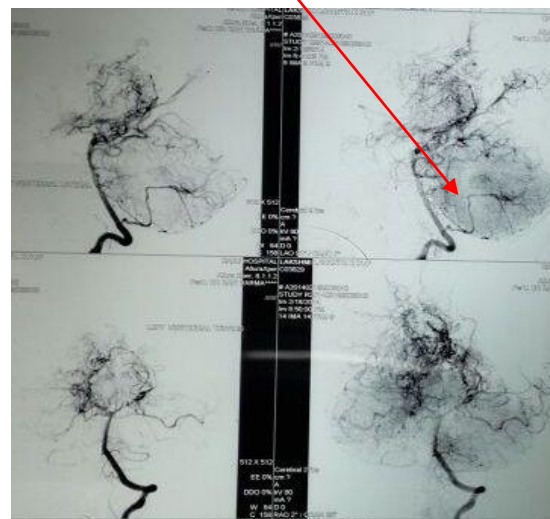
MR Angiogram showing severe stenosis in the distal Internal Carotid Artery in Supraclinoid segment



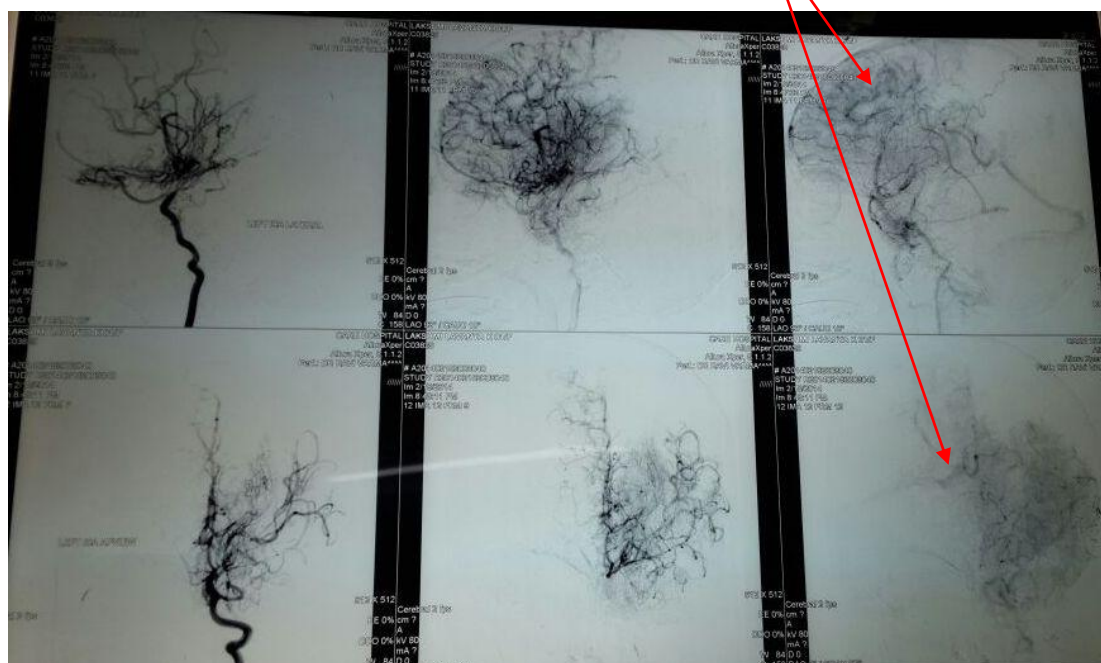
Cerebral Angiogram of Right Carotid injection showing supraclinoid occlusion with collaterals



Cerebral Angiogram of Vertebro Basilar injection showing multiple collaterals



“Puff of Smoke” appearance



### III. DISCUSSION

Moyamoya is Japanese word for 'puff of smoke' and describes the appearance of the resultant network of abnormal small collateral vessels seen on angiography. There is a familial form (about 15% of patients) which links to a gene on chromosome 17q25

- although the exact underlying cause remains unknown.

Pathologically there is fibrocellular intimal thickening, smooth muscle cell proliferation and increased elastin accumulation resulting in the stenosis of suprasellar

intracranial internal carotid arteries and also numerous perforating & anastomosing branches around the circle of Willis or in the peripheral vessels.

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. Clinical features include transient ischaemic attacks (TIAs) (more common with children) and stroke (haemorrhagic - more typical in adults). They include headache, hemiparesis, seizures, disturbed consciousness, speech deficits (aphasia), sensory and cognitive impairments, involuntary movements and vision.

The gold standard test is radio-imaging. Findings suggestive of the diagnosis of moyamoya disease on CT scanning or magnetic resonance angiography (MRA) include: Stenosis or occlusion at the terminal portion of the internal carotid artery or the proximal portion of the anterior or middle cerebral arteries. Abnormal vascular networks in the vicinity of the occluded or stenosed areas. No role of antiplatelet drugs, anticoagulants. Surgical procedures such as EMS or EDAS are useful in the management of Moya Moya disease.

#### **IV. CONCLUSION**

Moya Moya Disease is a rare disease which is prevalent in Asian population. Surgery improves the prognosis for patients, however the prognosis is poor. It is a rare but treatable disorder.

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