

## Moya Moya Disease : An interesting case of recurrent young stroke

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### ABSTRACT

Moyamoya disease (MMD) is a unique chronic progressive cerebrovascular disease characterized by bilateral stenosis of the arteries around the circle of willis with prominent arterial collateral circulation. This leads to a classical appearance of puff of smoke on angiography, hence the name. With the highest reported incidence among Japanese population it has been under reported in other parts of the world. Here we report a case of a young 14 year old Indian girl child who presented to the emergency medicine department with disorientation, aphasia, right sided weakness and numbness, blurring of vision, emotional instability since 2 days and with past history of similar complaints with complete recovery 4 years back. Imaging modality confirmed Moyamoya disease after we ruled out other causes of secondary moyamoya.

### Introduction :

Acute Ischemic Stroke (AIS) is one of the significant cause of morbidity and mortality in children and young adults although more common in older ones. Incidence of acute ischemic stroke (AIS) in young adults is as high as 22.8/ 100,000 people per year with male preponderance. The etiology of AIS in children and young adults differ from those typical in elderly- it includes congenital and acquired heart problems, hematologic conditions, vasculopathies (inflammatory and non inflammatory), metabolic disorders, and drug ingestion. The importance of this case is to find the etiology in a case of AIS for better neurological prognosis. Proper evaluation, early diagnosis and accordingly effective treatment can help in reducing the morbidity and mortality in these patients.

We reported a rare case presenting as acute ischemic stroke.

### Case Report :

A 14 year old female child brought to the medicine casualty with complaints of weakness of right half of body, emotional instability, aphasia, drowsiness since 1 day. There was no history of trauma, head injury, fever, convulsions. There was no history

suggestive of haemoglobinopathies, no history of joint pain, blood transfusion. She had a history of similar episode of stroke (AIS) of right half of body 4 years back which recovered over a period of 1 month without residual neurological deficit. No history of delayed milestones or any other medical illness was present. Other family members were not having similar illness.

On examination patient was conscious but disoriented. There was no obvious bony abnormality or skin lesions. She was afebrile with pulse of 94/min regular, BP 100/60 and RR of 16/min. There were no signs of meningeal irritation. Her spine examination was normal. On neurological examination, she was having global aphasia. On cranial nerve examination there was no evidence of papilloedema. Right sided supranuclear seventh nerve palsy was present. Her muscle nutrition was normal, tone was exaggerated and the power of muscles acting on all the joint of right side was 2/5. Her sensory system could not be tested. Her deep tendon reflexes were exaggerated and planter reflex was extensor on right side. Her other systems examination were within normal limit.

Her laboratory investigations like CBC, KFT, LFT, serum electrolytes, peripheral smear, coagulation profile, serum lipid profile, serum homocysteine, antinuclear antibody test, cANCA, pANCA shows no abnormality. Her HIV testing was non reactive. Her echocardiography examination was normal. Her MRI brain was done whose imaging was as:

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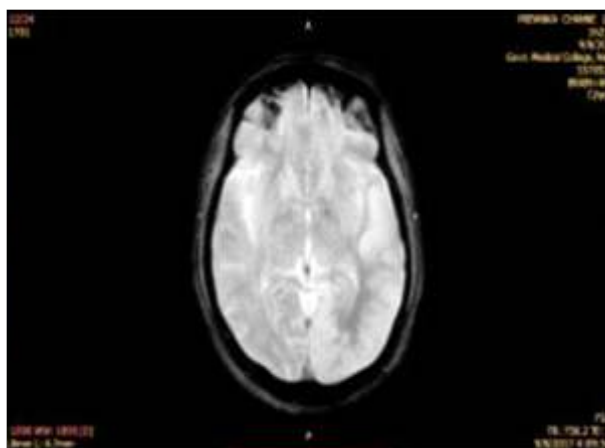
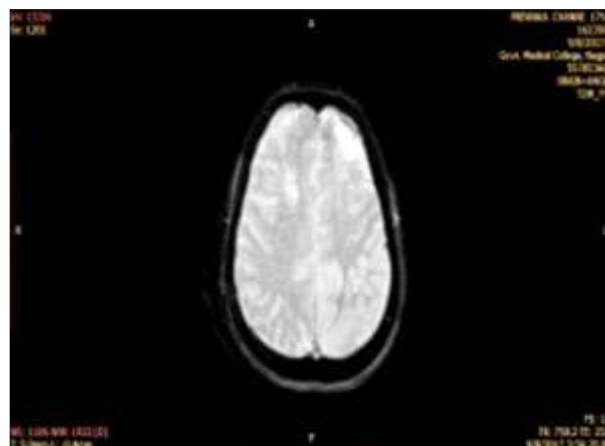
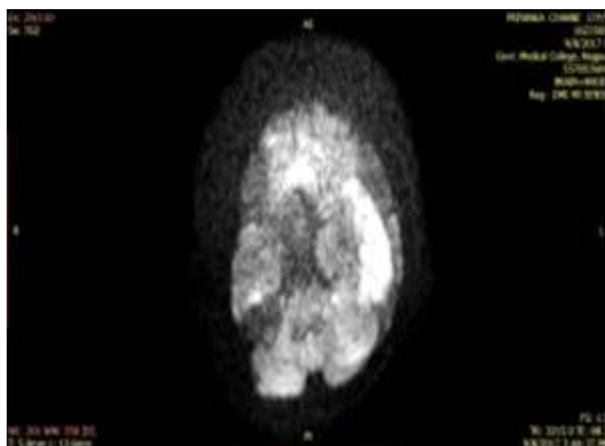
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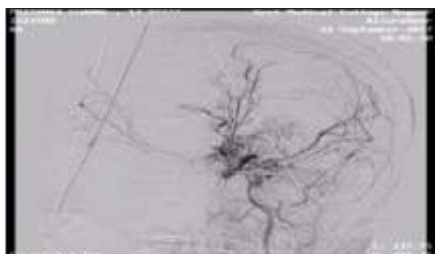
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Showing large acute infarct in left PCA and MCA territories.

Multiple chronic infarcts involving areas supplied by left MCA,ACA arteries.

Then a digital subtraction angiography was done which revealed narrowing of supraclinoid segment of bilateral ICA with abrupt cut-off and non-visualization of right MCA, bilateral ACA, faint visualization of left MCA, PCA; development of collaterals giving puff of smoke appearance in MCA, ACA and PCA territories f/s/o Moya moya disease.



Based on the above findings diagnosis of MOYAMOYA disease stage 4 had been made. Patient was stabilized during acute phase with hydration and started on aspirin 75 mg once daily. She was started on physiotherapy. After 7 days of hospitalization she was discharged on t aspirin 75 mg once daily while explaining future course of illness to relative.

#### Discussion :

Moyamoya disease is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels seen on imaging particularly cerebral angiography<sup>1</sup>. Moyamoya is a Japanese word for “puff of smoke”. Patient with angiographic appearance of moyamoya but no known risk factors are considered to have moyamoya disease while those with well recognized risk factors such as intracranial infections, atherosclerosis, cranial irradiation,

haemoglobinopathies (such as sickle cell disease etc.), vasculitis and autoimmune conditions, connective tissue disorders like neurofibromatosis, chromosomal disorders like Down's syndrome, cardiovascular such as congenital heart diseases etc. are classified as secondary moyamoya syndrome<sup>2</sup>.

The clinical manifestation of moyamoya is variable which include transient ischemic attack, ischemic stroke, hemorrhagic stroke, and epilepsy. Cerebral angiography is the gold standard imaging modality to diagnose this condition. However noninvasive imaging study such as digital subtraction angiography (DSA), MR Angiography and CT angiography are more commonly performed. Depending upon the angiographic progression of the disease it is classified into SUZUKI STAGES<sup>1,3</sup>:

- Stage 1 : narrowing of carotid fork only
- Stage 2 : initiation of basal moyamoya with dilatation of all main cerebral arteries
- Stage 3 : intensification of moyamoya together with reduction of flow in the middle and anterior cerebral artery
- Stage 4 : minimization of moyamoya vessels; proximal PCA gets involved
- Stage 5 : reduction of moyamoya and absence of all main cerebral arteries
- Stage 6 : disappearance of moyamoya vessels; the cerebral circulation is supplied only by the external carotid artery.

Management of the disease is divided into two parts i.e. acute management during episode and secondary prevention. Acute treatment is mainly symptomatic and directed towards reducing intracranial pressure, improving cerebral blood flow, controlling seizures and minimizing the

precipitating events such as crying and hyperventilation. During ischemic stroke thrombolysis is generally not indicated in view of intraparenchymal bleed through collaterals. Antiplatelets therapy is given but anticoagulation is considered risky in view of the possibility of cerebral hemorrhage. Secondary prevention aims at preventing further deterioration. It includes direct or indirect revascularisation surgeries such as transplantation of a vascular muscle flap, omentum, or pedicle containing the superficial temporal artery to the pial surface of the frontal lobe temporal pial synangiosis with the idea of creating neovascularization of the cortical convexity<sup>4</sup>. However in spite of all these, treatment of moyamoya is far from satisfactory.

#### **Conclusion :**

In its natural course Moyamoya disease leads to repeated AIS; early diagnosis and treatment would lead to better neurological outcome. This case highlights the importance of considering this disease as one of the differentials while dealing with the young children and also patients with recurrent stroke who are in their third/fourth decade of life

**Sources of Support :** Nil

**Conflict of Interest :** None

#### **References :**

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