

Case Report

MOYAMOYA DISEASE- A RARE CASE REPORT OF VANISHING CEREBRAL VESSELS

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ABSTRACT

Moyamoya disease is a rare cerebrovascular disease caused by progressive stenosis of distal internal carotid artery and their major branches. Moyamoya means puffy, hazy, obscure or vague, expressing the smoky look of vascular collateral network on angiography. The dilated and fragile basal collateral circulation display "PUFF OF SMOKE" appearance, thus called moyamoya vessels. Transient ischemic attack occurred most often during the first year of life, there after decreased intellectual deterioration and neurological deficit increase with time. Its unique features are 2:1 female preponderance and its peak incidence in two age groups- Early childhood and adults in their 40s. Most patients including my patient presented with complain of hypertension, seizures, hemorrhagic strokes, hemeperepsis and episodic headache.

Keywords: *Moyamoya Disease, Stroke, Seizures*

INTRODUCTION

Suzuki and Takaku first described moyamoya in 1969 (Jiro and Akira, 1969). Moyamoya disease is a poorly understood vascular disease primarily affecting individuals of asian descent with the largest peak at the age of 5 years. Moyamoya disease most commonly presents with seizures, stroke and episodic headache and is an important cause of cerebral stroke in children.

CASES

Reporting a case of 7 yr old male patient who came with complain of headache and vomiting since two months and with sudden onset of weakness in right sided upper and lower limb and right sided facial twitching with history of fall in the morning, vitals were unstable and blood pressure was high(140/90 mm hg). On CNS examination tone and power were less.

MRI brain with angiography was done and report revealed- Few small focal abnormal signal intensity areas showing restricted diffusion in left high frontal and high parietal region suggestive of small acute infarcts. Ventricle, cerebral sulci and cistern appear prominent suggestive of cerebro-cortical atrophy. Multiple focal abnormal signal intensity areas hyperintense on T2/FLAIR in bilateral periventricular white matter suggestive of chronic ischemic changes. Complete short segment occlusion involving supraclinoid segment of bilateral internal carotid artery. Multiple tortuous collateral arteries at base of skull, in bilateral cerebral hemispheres and posterior circulation. Bilateral PCOM appears dilated; Findings are suggestive of- Moyamoya disease.

DISCUSSION

Moyamoya disease is thought to affect the primary individual of asian descent. Age at onset demonstrated with largest peak at age of 5 years. Moyamoya disease is poorly understood vascular disease characterized by compensatory collateralization that follows arterial occlusion at the base of the brain.

Although predominantly a disease of children and females, moyamoya disease can occur at any age. Originally moyamoya disease was thought to develop chronically over months to years however it can occur rapidly within two months following subarachnoid hemorrhages. Cerebral angiography is the gold standard however it is not mandatory for diagnosis. MRI and MRA also show typical findings with more than 1.5T magnet. A novel grading system for MRA as been developed based on severity of occlusive changes of ICA, ACA, MCA and PCA and visibility of distal branches. MRA score correlates with the

Case Report

six stages of cerebral angiography. MRA grade-I correlates with angiography stages 1 and 2 and MRA stages 2 and 3 correlates with angiography stages 3 and 4 and 5 mainly.

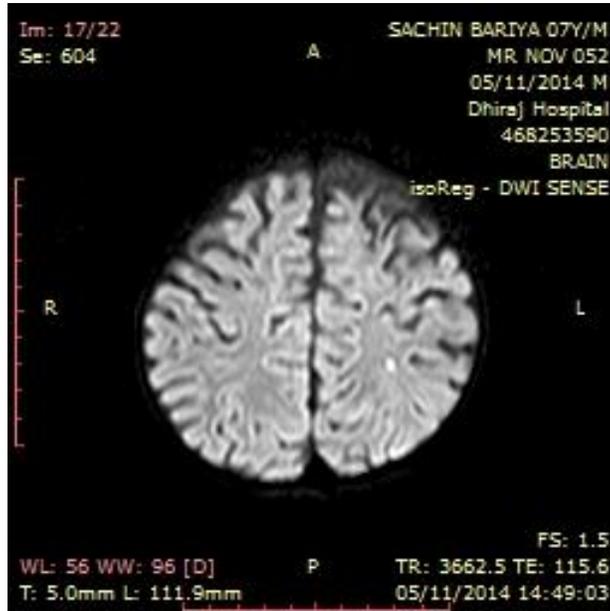


Figure 1: Few small focal abnormal signal intensity areas showing restricted diffusion are noted in left high frontal and high parietal region suggestive of small acute infarcts

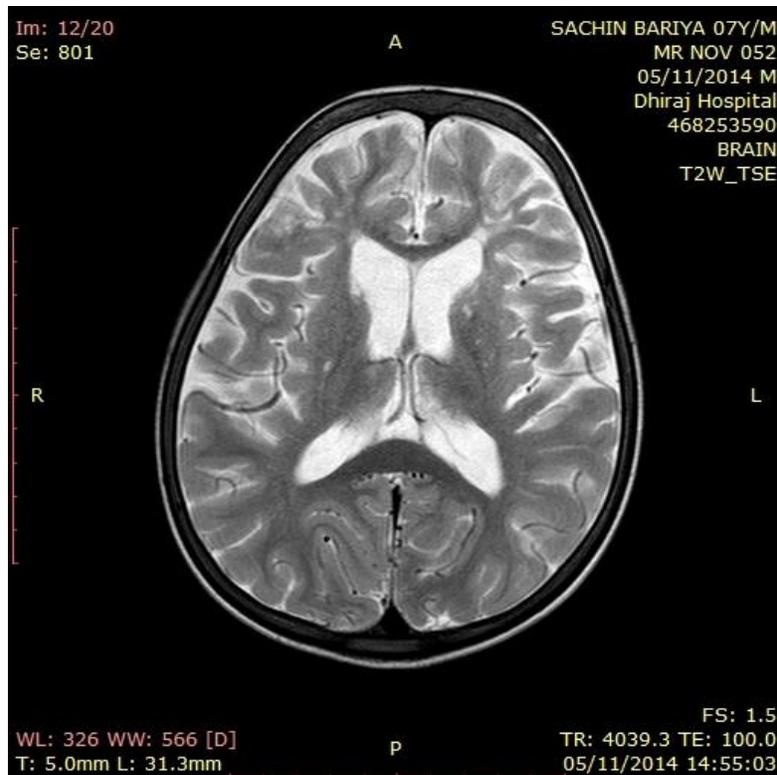


Figure 2: Ventricle, cerebral sulci and cistern appear prominent suggestive of cerebral cortical atrophy. Multiple focal abnormal signal intensity areas hyperintense on T2/FLAIR noted in bilateral periventricular white matter suggestive of chronic ischemic changes

Case Report



Figure 3: Complete short segment occlusion is noted involving bilateral internal carotid artery. Multiple tortuous collateral arteries are noted at base of skull, bilateral cerebral hemispheres and posterior circulation. Bilateral PCOM appears dilated

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