

## CRAO in Moyamoya Disease-The Puff of Smoke That Can Blind the Eye

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**Abstract:** Moyamoya (Japanese word meaning; moya-tiny) disease [MMD] is a rare idiopathic progressive occlusive cerebro-vascular disease of a terminal portion of internal carotid artery or a proximal portion of the anterior cerebral arteries and the middle cerebral arteries. These abnormal tiny vascular network seen in the base of the brain, produce 'puff of smoke' appearance on Magnetic Resonance angiography. Although MMD commonly is seen among Japanese population, it is occasionally reported in Indian population. We report a case of unilateral central retinal artery occlusion(CRAO) in a young male who was later diagnosed as a case of MMD on further radiological evaluation.

**Keywords:** Moyamoya disease, stroke, central artery occlusion.

### I. Introduction

Takeuchi and Shimizu first described the disease entity in 1957<sup>1</sup> and Suzuki and Takaku named the disease after Japanese term "Moyamoya" in 1969.<sup>2</sup> The appearance of vascular abnormalities based on brain angiography was named as 'a hazy cloud like a puff of cigarette smoke'.<sup>2</sup> Commonly seen among Japanese population, the prevalence and incidence of the disorder there has been reported to be 3.16 cases and 0.35 case per 100,000 people, respectively.<sup>3</sup> It is more commonly seen in females.( 1.8:1) The onset of MMD can be bimodal, with a predilection for ischemic cerebro-vascular events in childhood and hemorrhagic strokes in adults.<sup>4</sup>

We report a rare case of MMD involving posterior cerebral artery with central retinal artery occlusion in an adult male from south part of India.

### II. Case report

A 45y old male presented with painless sudden onset loss of vision in right eye of 1 day duration. Diminution of vision was associated with weakness of left side of the body, right sided headache. On ocular examination, his best corrected vision was counting finger close to face in his right eye and 6/6 in his left eye. He had normal anterior segment in both his eyes. Right eye showed relative apparent defect on swinging torch light test. Fundus examination showed retinal whitening, cherry red spot at macula in the right eye suggestive of central retinal artery occlusion. He was treated with ocular massage, paracentesis and systemic acetazolamide tablets. His previous records showed that he had repeated episodes of stroke, 2 episodes of transient ischemic attack. He was also a known hypertensive on treatment since past 3years. In view of the left sided facial weakness and headache the patient was immediately referred for neurological and cardiovascular evaluation. He underwent further tests which are follows.

Routine blood investigation revealed normal Complete Blood Counts, Erythrocyte sedimentation rate, random blood sugar, C-reactive protein, antinuclear antibody titres and his VDRL/ HIV tests were negative. Coagulation profile and chest X-ray were normal.

Echocardiography showed concentric left Ventricular Hypertrophy with an ejection factor of 56%. Carotid Doppler revealed an echogenic eccentric plaque in the right internal carotid artery causing 55 to 60% stenosis.

MRI brain and neck showed old right Middle Cerebral Artery infarct. Multiple acute infarcts were seen in the right middle cerebral artery territory and anterior cerebral artery. (Figure 1) Right internal carotid artery and middle cerebral artery occlusive disease was noticed. Old infarcts were also seen in the right temporal and parietal lobes.

A cerebral digital subtraction angiography revealed right sided supra-clinoid internal carotid artery occlusion, with the intracranial views showing multiple collaterals from anterior cerebral artery, with posterior cerebral artery branches supplying right middle cerebral artery territory which appeared like a "puff of smoke", suggestive of a diagnosis of MMD. (Figure 2) He was managed with systemic medication like statins, low

molecular weight heparin, anti-platelet drugs, and other supportive measures. Patient later underwent neurosurgical intervention for intracranial extra-cranial bypass procedure.

Two months after initial presentation his ocular examination revealed a BCVA of counting fingers 3 meters in his right eye and 6/6 in his left eye. IOP was within normal limits in both the eyes. Right eye had a relative afferent pupillary defect; anterior segment did not reveal any iris or angle neovascularisation. Fundus evaluation showed a pale disc with arteriolar attenuation in the right eye consistent with old CRAO. (Figure 3) Extra ocular movements were normal in both the eyes. As the patient had undergone an intracranial extra-cranial bypass procedure, Fundus fluorescein angiography was done to study the retinal circulation. It revealed a delayed retinal arterial filling in the right eye, as compared to the left eye with normal choroidal fluorescence indicating an old CRAO in the right eye. No neovascularisation was seen in the right eye. (Figure 4).

### III. Discussion

Moyamoya disease is commonly seen in the Japanese population but it can also occur in other populations. The condition is believed to be hereditary and linked to q25.3, on chromosome 17.<sup>7</sup> Disease can present in paediatric or in adult age group, with varied clinical presentations. Children present with repeated ischemic events whereas haemorrhagic events are more common in adults. The symptoms and clinical course vary widely; some patient may be asymptomatic while in others it can cause transient neurological events or cause severe neurologic deficits. It causes progressive occlusion of the circle of Willis, primarily involving internal carotid artery, leading to abnormal dilated collateral vessels, which look like “a puff of cigarette smoke” (Moyamoya) on cerebral angiography.

Ischemic stroke, brain haemorrhage, seizures and death are the major complications of Moyamoya disease. In persons with moyamoya disease, the death rate (usually from brain hemorrhage) is somewhere between 5-10%. Young adults with moyamoya typically present with recurrent attacks of headache, transient ischemia and stroke as was seen in our patient.

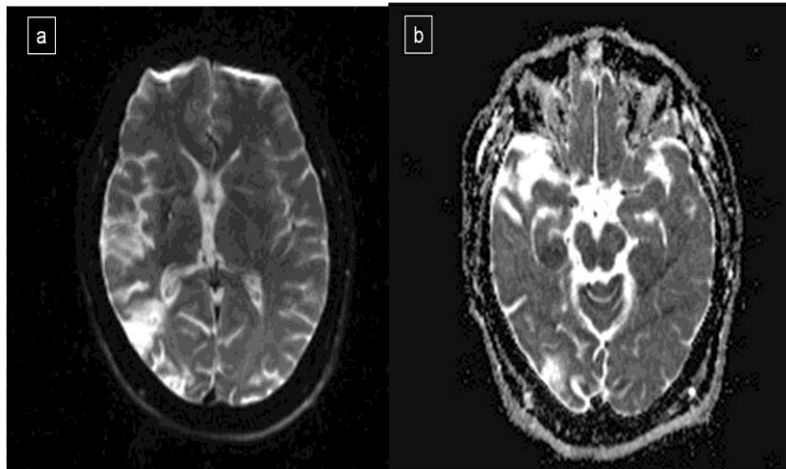
The ocular manifestations of MMD are rare but their presentation varies. It may present with vague symptoms like amaurosis fugax, diplopia or with severe loss of vision or hemianopia.<sup>5</sup> Ocular perfusion anomalies in adult MMD have been reported only in 3 cases in the literature. Few cases of hemi CRAO, ischemic optic neuropathy, chorio-retinal atrophic patches, acute CRAO<sup>8</sup> and central retinal vein occlusion<sup>9</sup> have been reported in patients with MMD. Although vascular abnormalities in MMD predominate in the anterior and middle cerebral arteries, the posterior cerebral artery has been found to be involved in the course of the disease in 50% of patients.<sup>6</sup> which was a finding noted in our case. The symptom of pain should alert possibility of ocular ischemic syndrome, seen in 40% of peoples<sup>10</sup> which is typically, characterized by dull ache over the eye or brow due to ischemia to the anterior segment structures, also known as ocular angina. Our case presented with the clinical features of acute CRAO which on further investigation was diagnosed to have MMD. CRAO is rare in patients who are younger than 50 years of age and in the younger patients, emboli from the heart are a major cause. The highlight of this case is that Moyamoya Disease is a rare disease in this part of the world which was the cause of Central Retinal Artery Occlusion in this young adult male patient hence when evaluating a case of CRAO in a young patient we may need to consider rare causes like Moyamoya disease even in Indian population, especially when the patient has a repeated history of stroke, headache, transient ischemic attacks.

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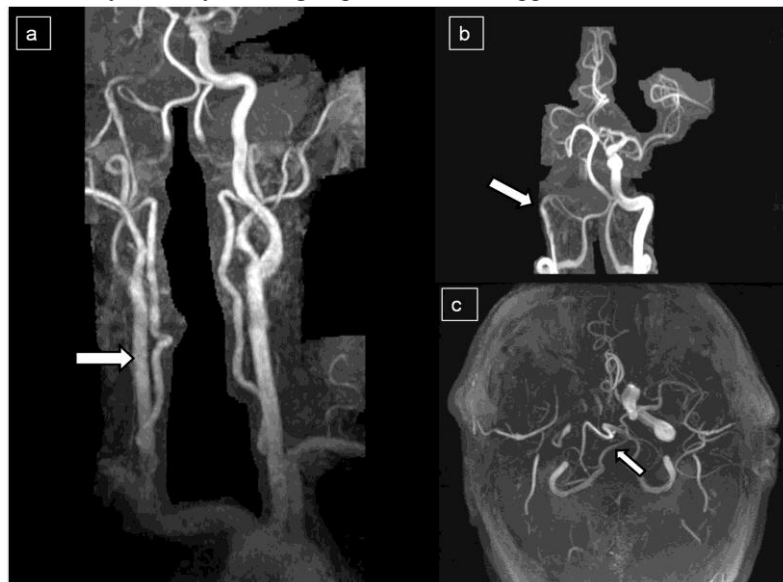
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**Legends:**

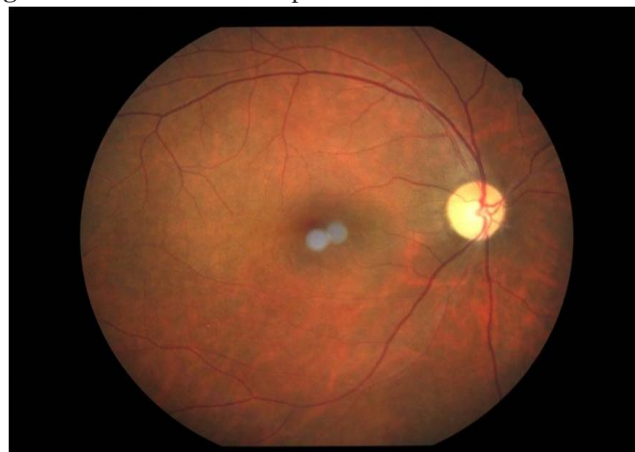
**Figure 1:** MRI brain showing old right Middle Cerebral Artery infarct (a) and multiple acute infarcts in right Anterior Cerebral Artery territory (b).



**Figure 2:** MR- Digital subtraction angiography showing right side supraclinoid internal carotid artery occlusion (a) with multiple collaterals from anterior cerebral artery and posterior cerebral artery branches supplying right middle cerebral artery territory creating a ‘puff of smoke’ appearance in intracranial views. (b,c)



**Figure 3:** Color Fundus OD-pale disc with arteriolar attenuation,



**Figure 4:** OD -(A): Choroid filling at 17sec, (b): Arterial filling 19 sec, (c): Lamellar flow 26 sec , (d-e):A-V transit 26 sec to 32 sec; OS (4g-h): normal study

