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Moyamoya disease: A case report

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Abstract

Moyamoya disease is a rare cause for hemorrhagic stroke in adults. The authors present the case of a 39 years old man, who presented with features of intracerebral hemorrhage secondary to Moyamoya disease. He was a known case of diabetes mellitus, hypertension and had a past history of ischemic stroke. Left temporoparietal craniotomy and encephaloduroarteriomyosynangiosis (EDAM'S) was performed for the same. Patient got discharged on fifth post-operative day without any complication. The etiopathogeneses, clinical features, and management are discussed in this paper.

Key words: Moyamoya disease, encephalodurangiomyongiosis (EDAM'S), digital subtraction angiography (DSA), stroke, headache

Introduction

Moyamoya disease is a chronic cerebrovascular disorder characterized by an initial obliterative vaso-occlusive phase followed by a vaso-proliferative phase. The progressive occlusion of supraclinoid intracranial Internal Carotid Artery (ICA) and its proximal branches, mainly middle cerebral artery and anterior cerebral artery is seen in the initial phase. As a compensatory phenomenon and collaterals develop from the basal perforators (Duan et al., 2012)These collateral vessels appear as "smoky puffs" in carotid angiography. Though it was first described by Takeuchi and Shimizu in 1957 (Takeuchi & Shimizu, 1957) and later in 1969, Suzuki and Takaku called it as Moyamoya (means puff of smoke in Japanese) (Suzuki & Takaku, 1969).

Case report

A 39 years old male patient presented to neurosurgery department with the history of severe headache for one week. The headache was holocranial in nature. There was a history of associated vomiting and giddiness with blurring of vision. There was no history of any new onset of limb weakness, transient ischemic attack (TIA) or sudden loss of consciousness in last two years.

Patient had the past history of right Cerebrovascular Accident (CVA) four years back. He also had bilateral pyelonephritis with papillary necrosis with Enterococcal Sepsis and Candidemia. Bilateral double J (D-J) stenting was done four years back. Patient is a known case of Type-2 diabetes mellitus and hypertensive for four years and on regular treatment.

Patient was conscious, alert, and oriented. His Glasgow Coma Scale (GCS) was 15 (E4 V5 M6). His vital signs were stable; heart rate was 80/min, blood pressure was 140/100 mmHg, respiratory rate was 20 breaths/min, and temperature was 98.6^0 F. Right side pupil was measuring two mm and was reacting to light. He had intraocular lens in the left side. Higher mental function was normal and had mild slurring of speech. Fundoscopic examination of the eye revealed papilloedema. Muscle tone was normal with power 4/5 in right upper and lower limb and pronator drift was positive. However, there was no sensory deficits and deep tendon reflex was 2+ with no cerebellar signs.

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Hematological parameters (renal function test, blood glucose levels, serum electrolytes and blood counts) were within the normal limit. CT brain was suggestive of intraventricular hemorrhage in bilateral lateral ventricles and third ventricle with dilatation of body and temporal horn of left lateral ventricle (Figure 1).

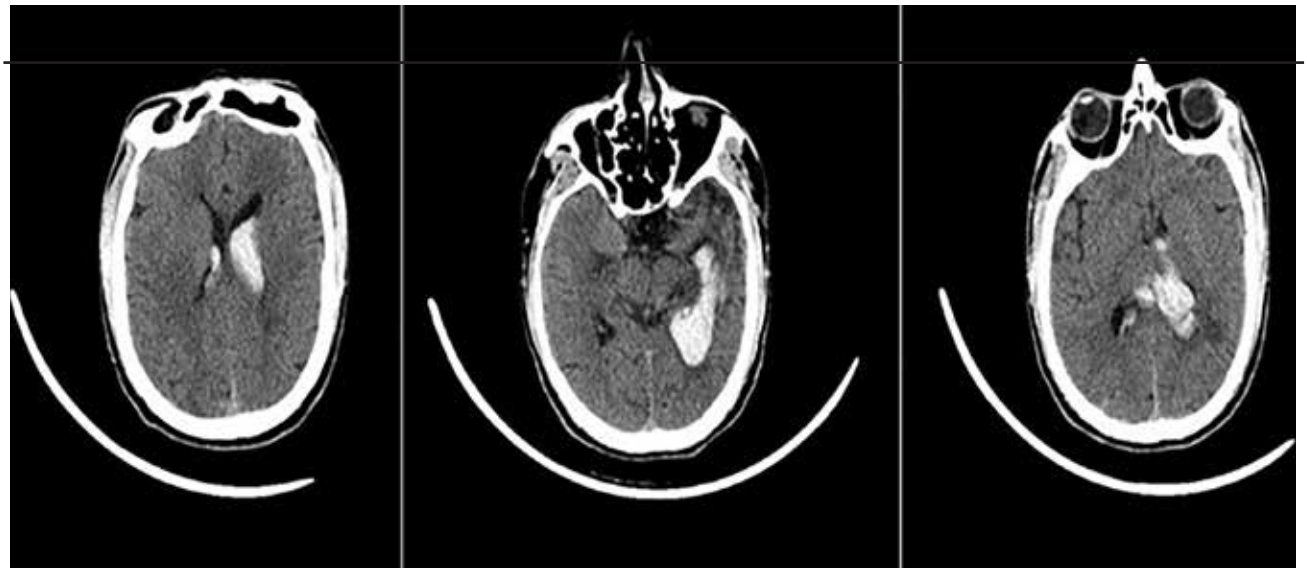

Figure 1: CT scan showing presence of bleed in the lateral ventricle – body, temporal horn and atria

Digital subtraction angiography was suggestive of stenosis of anterior cerebral artery and middle cerebral artery (Figures 2, 3 and 4).

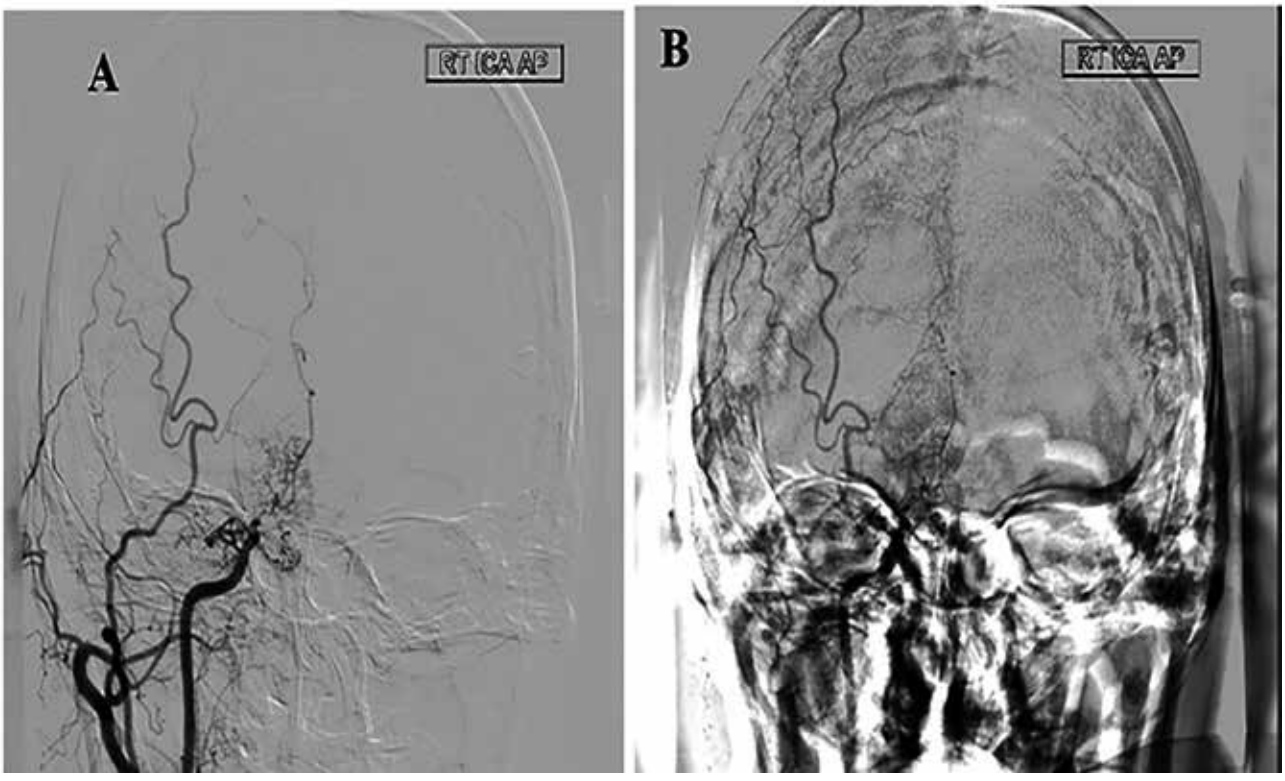

Figure 2: A: Right ICA AP injection showing stenosis of supraclinoid ICA & Moyamoya vessels. B: Right ECA injection showing collaterals from ECA

ICA: Internal carotid artery, ECA: External carotid artery

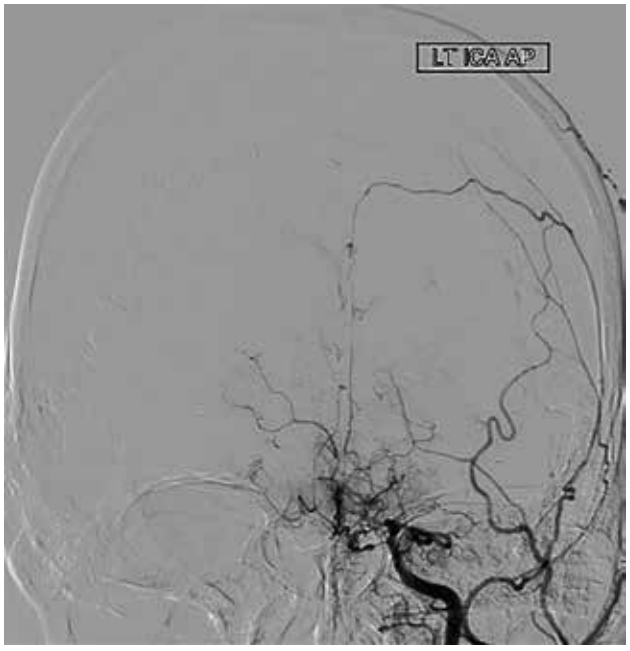

Figure 3: Left ICA injection showing a) occlusion of supraclinoid ICA and b) blush of collaterals from the basal perforators

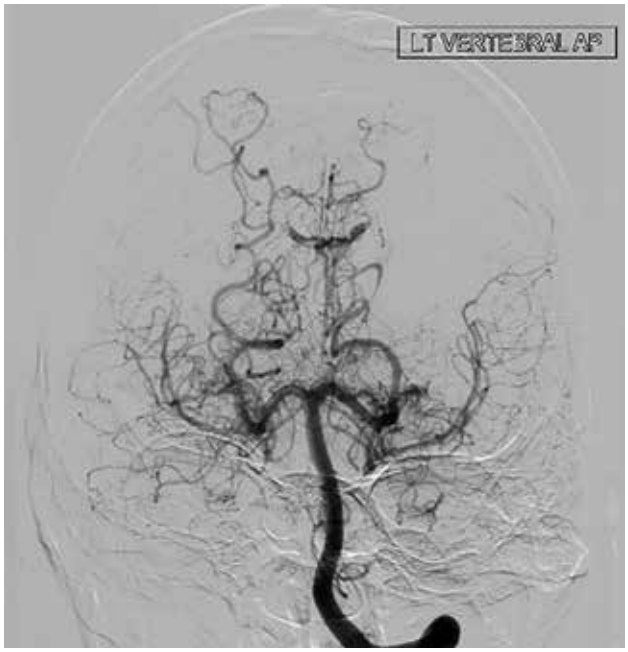

Figure 4: Vertebral injection showing diffuse blush of perforators

Left temporoparietal craniotomy and EDAM'S was performed. Post-operative period was uneventful.

Discussion

Moyamoya disease is a rare cause for stroke characterized by progressive stenosis of internal carotid artery and formation of a collateral network of basal perforators. These basal collaterals mimic a "puff of smoke" from which the name Moyamoya was derived (Gosalakkal, 2002).

Moyamoya disease was initially thought to be only found among the Japanese and the oriental population. But of later, several reports from India and western countries are being published [(Duan et al., 2012) (Chiu, Shedden, Bratina, & Grotta, 1998); (Yilmaz, Pritz, Bruno, Lopez-Yunez, & Biller, 2001); and (Uchino, Johnston, Becker, & Tirschwell, 2005)]. Japanese population has a higher incidence of pediatric Moyamoya whereas the western literature cites a higher incidence of adult Moyamoya.

Moyamoya patients present either with ischemic stroke or with intracerebral bleed. The ischemic stroke is more common among children. The bleeding is due to rupture of delicate collaterals or rupture of small aneurysms seen in these collaterals. Usually, adults are vulnerable for bleeds. The clinical manifestations are headache, transient ischemic attack, infarct, hemorrhage, seizure, syncope [(Duan et al., 2012) and (Han et al., 2016)]. However, some of the patients remain asymptomatic (Duan et al., 2012) Among all the symptoms, ischemia is noted to be the most frequently occurring symptoms (Duan et al., 2012) and is the most common form of presentation among children.

Moyamoya usually affects bilateral carotid arteries than unilateral (Duan et al., 2012) It is still unclear if all patients with unilateral MMD progress to bilateral MMD. The etiopathology of the disease is unclear. MMD is often associated with neurofibromatosis, Down syndrome, Turner syndrome, sickle cell anemia, Wilms tumour, meningitis, and cranial irradiation. Some authors prefer to call such secondary cases as Moyamoya syndrome and the others as Moyamoya disease. The identified risk factors are hypertension, diabetes mellitus, dyslipidemia, hyperhomocysteinemia and carotid plaques (Han et al., 2016), female, family history (Duan et al., 2012) smoking, ischemic heart disease and hypertension (Jin et al., 2016). The pathogenesis of Moyamoya is not well established. One hypothesis states that the disease is associated with the growth of fibroblast which is responsible for the angiogenesis (Malek, Connors, Robertson, Folkman, & Scott, 1997). Another postulate is role of prostaglandin in promoting the thickening of cerebral blood vessels

leading to partial or complete occlusion (Yamamoto, M., Fukai, Matsushima, & Yamamoto, 1999).

The vascular changes are evident in imaging studies such as cerebral angiogram and High-Resolution Magnetic Resonance Imaging (HRMRI) showing intimal fibrous thickening, widening of the internal elastic lamina and thinning of the media and presence of collateral around the circle of Willis and small vessels of conglomerated networks in the pia mater (Gosalakkal, 2016). HRMRI is useful in diagnosing Moyamoya disease. HRMRI not only helps in diagnosis of Moyamoya, it also found to be useful in detecting atherosclerotic plaques along with Moyamoya progression [(Han et al., 2016) and (Yu et al., 2016)]. Still, DSA remains as the gold standard in diagnosing the disease. Occlusion levels are categorized in terms of Suzuki staging (Table 1).

Table 1:
Suzuki Stages of Moyamoya Disease (Takahashi, Fujiwara, & Suzuki, 1986)

Suzuki Stage	Angiographic Finding
I	Narrowing of carotid arteries
II	Initial appearance of Moyamoya vessels
III	Intensification of Moyamoya vessels
IV	Minimization of Moyamoya vessels
V	Reduction of Moyamoya vessels
VI	Disappearance of Moyamoya vessels

The effective treatment of Moyamoya is not yet proven. The surgical procedures performed to treat the disease among adult patients are direct bypass, indirect bypass and combined bypass. In direct bypass, a superficial temporal artery (STA) is anastomosed to middle cerebral artery. Direct method of revascularization is effective in patients with ischemic strokes. Indirect procedures which augment the blood supply to the deprived areas of cerebral tissue are Encephaloduroarteriosynangiosis (EDAS), Encephalomyosynangiosis (EMS) or Omental cerebral transposition. However, a recent meta-analysis shows that direct or combined bypass gives better results in terms of revascularization (Kim et al., 2016). Few patients would develop peri-operative complications such as symptomatic stroke, permanent neurologic deficits, and rarely death (Kim et al., 2016). Nevertheless, there is a high risk of stroke among non-surgically treated adult patients (Jin et al., 2016), (Lee et al., 2012) and surgery is the current standard of care for patients with proven Moyamoya disease.

The disease progresses without treatment. The patients with ischemic stroke remain with neurological deficits. Surgery helps to arrest the advancement of disease. Prognosis following surgery is good and majority may not have disabilities (Duan et al., 2012)

Our patient presented with features suggestive of hemorrhagic Moyamoya disease. The bleed in such patients is attributed to rupture of the fragile collateral or rupture from a small aneurysm in these collaterals. Role of surgery in Moyamoya disease is debatable. However, since our patient had a relapse, we decided to perform a revascularization procedure. He underwent indirect anastomosis (EDAMS) and uneventful recovery and is doing well at one year follow up.

Conclusion

Moyamoya is a rare form of stroke which can be treated surgically. Cerebral revascularization helps in preventing further progression of the disease and thereby improving the quality of life. Proper awareness and prompt diagnosis helps in early detection and management.

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