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Case Report

"Moyamoya Disease"–Puff of Smoke in Brain-Presenting as Stroke in Young"

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Abstract

Moyamoya disease is a rare, idiopathic, progressive occlusive condition involving cerebral vessels which includes mainly, stenosis of distal internal carotid arteries on both sides and anterior and middle cerebral arteries thereby leading to development of collateral vessels to compensate for the occlusion. The disease may develop in children and adults, but the clinical features differ. Moyamoya disease occurs predominantly in Japanese individuals but has been found in all races with varying age distributions and clinical manifestations. As a result, moyamoya disease has been under-recognized as a cause of ischemic and hemorrhagic strokes in the Indian subcontinent. At this time, there is no known cure, and existing treatment options are controversial. We here describe the case of a 42-year-old male presenting as recurrent left sided hemiparesis.

Introduction:

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. It deemed a progressive steno-occlusive disease at terminal portions of the bilateral internal carotid arteries with the development of "moyamoya vessels" as collateral channels of circulation. The appearance of these small, multiple vessels at the base of the brain on catheter angiography was originally described by the Japanese term moyamoya, which translates to "puff of smoke" [1]. It was first described in Japan by Takeuchi and Shimizu in 1957 [2]. It is a rare disease with reported incidence of 0.086 per 100,000 population [3]. 3Although the disease is most common in Japan, many subsequent cases have been reported elsewhere, including North America, Europe, and India [4-6]. There are nearly twice as many female patients as male patients [7-9]. Although moyamoya syndrome has the same angiographic appearance as moyamoya disease, it is associated with other medical conditions such as arteriosclerosis, autoimmune disease, down syndrome, head trauma, meningitis, sickle cell disease, neurofibromatosis Type 1, and previous radiation therapy [10-14]. Very few cases of moyamoya disease presenting in adulthood in non-Japanese individuals have been reported [15].

Case Report:

Mr. X, a 38 years old married, muslim, normotensive, non-diabetic, non-smoker, non-alcoholic businessman, hailing from Jashore presented with the history of recurrent strokes(one in June 2015 and then another in Sept 2016) over the last 2 years. He first developed sudden onset weakness in the left upper limb with pain in the medial aspect of his left arm, in June 2015. MRI of brain then showed multiple micro vascular ischemic foci in both Para ventricular deep white matter regions and right centrum semiovale, and he was started on antiplatelet and statin. He then discontinued his medications a year later, 20 days after which he developed the same left sided weakness along with memory deficits and behavioral changes. But there was no loss of consciousness, convulsion, any speech, and swallowing or sphincter disturbances. Repeat MRI brain revealed sub-acute infarcts in left cerebral hemisphere, with small infarcts and features of CNC vasculitis in both cerebral hemispheres. He was then advised DSA, which he deferred. A repeat MRI brain in December 2016 was reported as sub-acute hemorrhagic changes/? AVM in left frontal lobe with deep white matter micro vascular ischemic changes. He was again advised DSA for which he went to CMC, Vellore for further workup.

The patient denied any family history of stroke, seizure, or cancer apart from his two brothers died of heart attack. Socially, the patient was employed in a clerical job in his hometown. He did not smoke cigarettes, consume alcohol, or use illicit drugs.

On examination, Pulse was 84 beats/min, regular, BP 110/70mm of Hg on left upper limb on supine position. No pallor, icterus, cyanosis, clubbing or enlarged palpable lymph nodes, JVP not raised and all peripheral pulses were equally felt. There were no signs of liver diseases or neuro cutaneous markers. On CNS examination, higher functions: he was fully conscious, oriented, MMSE: 30/30. There was no mood or psychotic symptoms. Cranial nerves: 2nd: pupils bilaterally reactive to light and accommodation, visual fields normal, visual acuity 6/6 in both eyes, fundus normal in both eyes. 3rd to 12th cranial nerves were intact. Motor function revealed normal bulk and tone in all four limbs. Muscle power revealed good neck flexion and extension with good hand grip bilaterally. Wrist (flexion & extension), elbow (flexion & extension), shoulder (abduction, adduction, flexion, extension) muscle powers were 5/5 bilaterally. Muscle power of hips, knees, ankles and EHL were 5/5 except power of left hip flexion, left knee flexion and left knee extension were 4+/5. There were no involuntary movements. On sensory system examination all modalities of sensation including cortical sensations were normal. Superficial abdominal reflexes were present in all quadrants. On DTR examination, biceps, triceps, supinator, knee and ankle jerks were (++) bilaterally with plantar down going on both side. Gait was normal. There were neither any cerebellar dysfunctions nor any signs of meningeal irritations. Spine, skull and examination of other system revealed no abnormalities.

On investigation: Hb 13.6gm%, PCV 39.2%, total WBC including differentials were in normal limit, Platelets 205000/cumm, ESR 27mm in 1st hr. Extensive thrombotic workup were done revealing normal PT, APTT, TT, fibrinogen, D-Dimer, Factor VII, Protein C, Protein S,AT III, Homocysteine , APCR. DRVVT for lupus anti-coagulant, ANA, Anti HIV, HBsAg, and Anti HCV were negative. VDRL was non-reactive. RBS, LFT's and RFT's were inconclusive. CSF examination revealed total 2 cell/HPF (Lymphocyte), CSG glucose 62mg/dl and CSF protein was 34mg/dl [Figures 1-5].

Repeat MRI with contrast was done in CMC; Vellore reported a chronic infarct in left frontal lobe, lacunar infarct in deep white matter of both frontal lobes in anterior watershed and internal watershed territories and chronic ischemic lesions in the right caudate nucleus head. There was hyper intensity over the sulci of right and left hemisphere with collaterals showing "ivy" sign. The cavernous ICAs appeared attenuated bilaterally, with loss of flow void being seen in the petrous segment of left ICA and mild wall thickening seen in the petrous segment of right ICA. A CT angiogram is suggested for further evaluation of intracranial vessels, aortic arch and neck vessels. Later cerebral angiogram using right common femoral artery retrograde puncture access was done which showed total occlusion of left ICA and occlusion of right supraclinoid ICA just above the origin of posterior communicating arteries and narrowing of proximal right ACA, left MCA and ACA with numerous small moya-moya collaterals from right ICA and left ICA branches. The vertebra-basilar arterial systems were normal. He was discharged at stable condition with aspirin 75mg OD and levetiracetam 750 mg bid and advised for follow up after 6 months. During his follow up on Dec, 2019 repeat MRI brain revealed sequel of prior hemorrhagic infarct in left middle frontal lobe gyrus, chronic infarcts in right high fronto-parietal lobe in vascular watershed zone. Now he is doing well with the above mentioned medications.

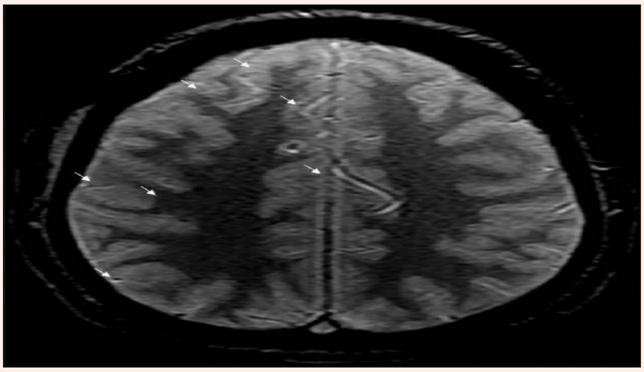

Figure 1: Arrows showing Hyper Intensity over the Sulci of Right and Left Hemisphere with Collaterals showing "Ivy" sign.

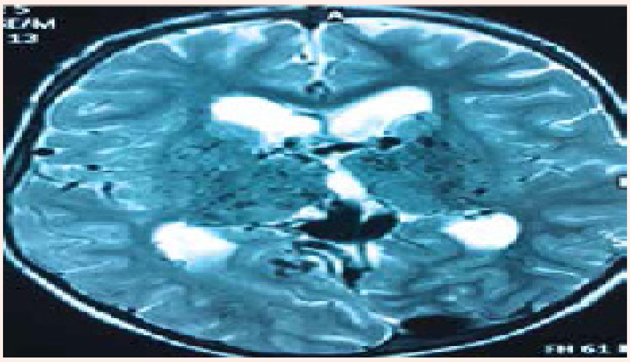

Figure 2: MRI of Brain T_2 image showed Multiple Flow Voids in the Bilateral Basal Ganglia.

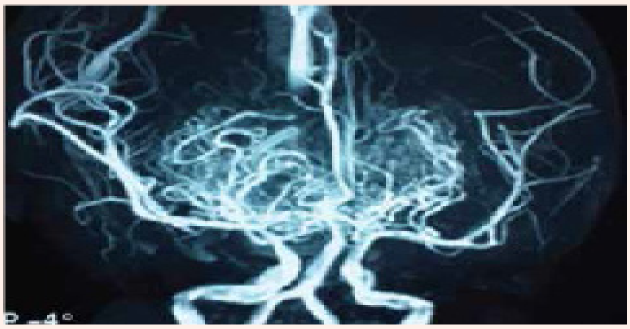

Figure 3: MR Angiogram (AP view) shows Stenosis of Bilateral Internal Carotid Arteries.

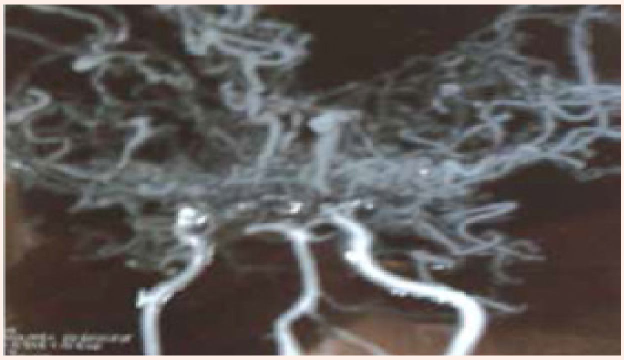

Figure 4: MR angiogram (A-P view).

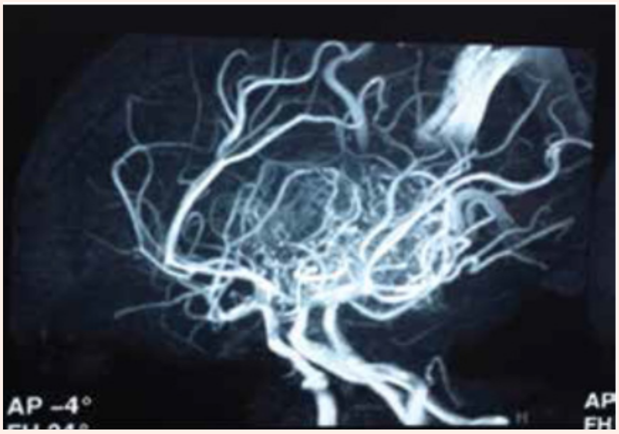

Figure 5: (Right Oblique View) shows Bilateral Stenosis of Internal Carotid Arteries with Typical Characteristic Moyamoya Vessels with "Puff of Smoke" Appearance.

Discussion:

Moyamoya disease is a rare idiopathic, slowly progressive vasculopathy characterized by irreversible stenosis of the arteries of Circle of Willis leading to cerebral hypo perfusion. The occlusive process stimulates the development of an extensive network of enlarged basal trans cortical and transdural collateral vessels.

Moyamoya disease though first reported in Japan in 1957, since then been reported worldwide. The disease has a particularly high incidence in Eastern Asia, especially in Japan. The overall prevalence rate of moyamoya disease in Japan in 1995 was 3.16 per lac with an incidence rate of 0.35 per lac. The male to female ratio was 1.8:1, and a family history of moyamoya was noted in 10% of cases. Family occurrence suggests that a genetic predisposition is present [16,17]. Unfortunately, to the best of our knowledge, more recent data are unavailable.

The process of narrowing of cerebral vessels seems to be a reaction of brain blood vessels to a wide variety of external stimuli, injuries, or genetic defects. Conditions such as sickle cell anemia, neurofibromatosis-1, Down syndrome, congenital heart defects, anti-phospholipid syndrome, renal artery stenosis, and thyroiditis have been found to be associated with moyamoya disease in the literature. But more than half of the adults seen with this disease have no cause for their moyamoya syndrome. Elevated levels of cellular retinoic acid-binding protein-I were found in the cerebrospinal fluid of patients with moyamoya disease [18]. High levels of hepatocyte growth factor, a known inducer of angiogenesis, have been found in the carotid fork and cerebrospinal fluid of patients with moyamoya disease [19]. Pathological changes in moyamoya patients include intimal thickening with fibrous tissue, abnormalities of internal lamina elastica, variable lipid deposition and virtual absence of inflammatory reaction in the blood vessel [20,21]. The process of blockage, once it begins, tends to continue despite any known medical management unless treated with surgery.

Clinically, the presentation of patients with moyamoya disease may include seizures, transient ischemic attacks, ischemic strokes, and hemorrhagic strokes [22-24]. Visual deficits, speech disturbance, migraine like headache, intellectual deterioration, cranial nerve palsies, and disturbance of gait can also be evident [25,26].

MRI not only reveals areas of infarctions but also allows direct visualization of these collateral vessels as multiple small flow voids at the base of brain and basal ganglia. MR angiography is used to confirm the diagnosis and to see the anatomy of the vessels involved. It typically reveals the narrowing and occlusion of proximal cerebral vessels and extensive collateral flow through the perforating vessels demonstrating the classic puff of smoke appearance [27].

Acute management is mainly symptomatic and directed toward reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures. Anticoagulant and antiplatelet agents have shown no remarkable benefit [28]. The same lack of obvious efficacy has been described for corticosteroids in moyamoya disease [29]. McLean et al., elucidated the use of verapamil hydrochloride to curtail the ischemic symptoms associated with moyamoya disease [30]. Revascularization procedures are currently performed to increase the perfusion to the hypoxic brain tissue. Direct revascularization techniques, which are typically used in adults, include the superficial temporal artery to middle cerebral artery bypass or the middle meningeal artery to middle cerebral artery bypass are commonly applied in children. The literature supports these procedures, and the long-term favorable outcome has been reported in terms of improvement in symptoms and positive angiographic follow-ups in all age groups.

Although Moyamoya is predominant in Japanese population but should not overlooked in other population. The patient who will fulfill the clinical characteristics, MR angiogram should be done to diagnose Moyamoya disease.

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