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Case Report

Moyamoya Disease in a 70 Year Old Nigerian Male: A Case Report

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Abstract: Moya-moya disease is a very rare disease characterized by a progressive vasculopathy leading to stenosis of the main intracranial arteries. Cerebral blood flow is usually impaired by constriction of the vessels with collateral circulation developing around the blocked vessels to compensate for the blockage. This may lead to an ischaemic stroke as observed on the index case. The aim of this case report is to highlight moyamoya disease as a rare cause of ischaemic stroke in an elderly male. We report a case of a 70 year old Nigerian male who has hypertension and diabetes mellitus, and presented with features of right hemispheric ischaemic stroke. The patient was investigated further with cerebral angiography and was found to have moyamoya disease. Angiography of the index case showed numerous abnormal hazy looking vessels arising from the level of the circle of willis and equivalent non filling in the posterior supratentorial and infratentorial spaces typical of moya moya pattern. He responded to medical therapy administered in the acute phase and subsequently followed-up at hospice care. Patient showed some clinical improvement with significant residual motor deficit on the left side of the body. The acute management was mainly symptomatic and directed towards reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures. The index patient was frail, unfit and therefore could not benefit from surgical revascularization procedure which is the main treatment option for moyamoya disease.

Keywords: Moyamoya Disease, Ischaemic Stroke, Cerebral Angiography and Revascularization

1. Introduction

Moyamoya disease is rare cause of ischaemic stroke and is characterized by a progressive vasculopathy leading to stenosis of the main intracranial arteries. Blood flow is blocked by constriction and thrombosis of the affected cerebral vessels. A collateral circulation develops around the blocked vessels to compensate for the blockage, but the collateral vessels are small and weak, and therefore prone to bleeding, aneurysm and thrombosis [1].

Moyamoya disease was initially described in 1957 as a "bilateral hypoplasia of the internal carotid arteries (Takeuchi and Shimizu 1957). The name of the disease comes from Japanese word and means *'puff of-smoke'. Moyamoya disease was later described in Japan by Takeuchi and Shimizu in 1963 as a* vaso-occlusive disease involving the internal carotid arteries and circle of Willis and was later named moya moya by Suzuki and Takaku in 1969 [2].

Suzuki and Takaku [3] classified the development of Moyamoya disease into 6 stages. According to this classification, many patients fall into stage 3. The stages include;

Stage 1: Narrowing of carotid fork.

Stage-2: Initiation of the "Moyamoya vessels", dilatation of the main intracerebral arteries.

Stage-3: Intensification of the "Moyamoya vessels", non-filling of the anterior and middle cerebral arteries.

Stage-4: Minimization of the "Moyamoya vessels", disappearance of the posterior cerebral artery.

Stage-5: Reduction of the "Moyamoya vessels", the main arteries arising from the internal carotid artery disappear.

Stage-6: Disappearance of the "Moyamoya vessels", the original Moyamoya vessels at the base of the brain are completely missing and only the collateral circulation from

the external carotid artery is seen.

The aetiopathogensis is not well understood but is believed to be multifactorial. Genetic factor has been implicated. Genetic background is frequently mentioned because in 10-15% of cases, the disease is found in other family members [2]. The proliferation of smooth muscle cells and fibrous tissue deposition in the walls of the moyamoya affected arteries has been found to be representative of the disease. These vessels are the ACA (anterior cerebral artery), MCA (middle cerebral artery), ICA (internal carotid artery) and posterior cerebral artery. The occlusion of the ICA is by far the commonest [4]. One hypothesis states that the disease is associated with fibroblast growth factor, which is responsible for the angiogenesis [5]. Another postulate is the role of prostaglandin in promoting the thickening of cerebral blood vessels [5].

There are conditions that are closely associated with moyamoya disease. Vascular lesions typical for moyamoya disease may also develop in patient with HCV infection, cryoglobulinemia, sickle cell disease, Down syndrome, neurofibromatosis and after radiotherapy of tumors of the optic chiasm [6]. Moyamoya mainly presents as ischaemic or haemorrhagic stroke. Following an ischaemic stroke, secondary bleeding may also occur. Commonly reported clinical features include headache, seizures, hemiparesis, dizziness, confusion and loss of conscious [6, 7].

Diagnosis can be confirmed via cerebral angiography. Studies have shown that Digital Subtraction Angiography remains the gold standard in diagnosing the disease. The vascular changes are evident on imaging studies such as CT 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. HRMRI are also useful in detecting atherosclerotic plaques along with Moyamoya progression [7]. Often nuclear medicine studies such as SPECT (single photon emission computerized tomography) are used to demonstrate the decreased blood and oxygen supply to areas of the brain involved with moyamoya disease [4].

The treatment for moyamoya disease is mainly revascularization surgery. Although, there are no effective medical therapies for the disease, Surgery remains the best available treatment option. Revascularization is thought to improve cerebral perfusion. Surgical procedures are classified into three categories; direct bypass which involves superficial temporal artery to Middle Cerebral Artery (STA-MCA) anastomosis, indirect bypass involves encephaloduroarteriosynangiosis (EDAS) and encephalomyosynangiosis (EMS), and combined bypassing [8].

The aim of this case report is to highlight moyamoya disease as a rare cause of ischaemic stroke in an elderly Nigerian male.

2. Case Presentation

Mr O. F is a 70 year old male who presented to our emergency room with a 10-day history of sudden onset headache and left sided hemiparesis with associated vomiting.

Eight days later, he had one episode of seizures characterized by jerking of all four limbs after transient stiffening, this lasted for about 3 minutes with postictal sleep and associated altered sensorium, there was however no neck stiffness, fever, prior head trauma, sleep deprivation or polypharmacy and also no family history of stroke, sudden death or substance abuse. He has hypertension and diabetes mellitus for more than 10 years, but not regular on medications and clinic visits. He had no family history of HTN, DM or cardiac disease. He occasionally takes alcohol but does not smoke cigarettes. He does not use recreational drugs and has no known drug /food allergies.

Physical Examination revealed an elderly man, unconscious and in respiratory distress (RR=32 breaths/min). He was afebrile, not pale, anicteric, acyanotic, not dehydrated and no pedal edema.

Central nervous System Examination showed a Glasgow coma score: 7/15 (E=2, V=1, M=4). Both pupils measured 2mm, poorly reactive with conjugate gaze deviated to the left. Cranial Nerve Deficits noted in: CN - IX & X (Poor Gag) and no neck stiffness. Tone and Reflexes were reduced in Left Upper and Lower Limb but were normal in Right Upper and Lower Limb. Patient moves the right side more than left when stimulated.

Pulse rate was 89b/m, normal volume, regular, synchronous with other peripheral pulses. Presence of arterial wall thickening, blood pressure was 164/98mmHg and JVP was not raised. Apex beat was located at 5LICS lateral to MCL, not heaving. HS: I, II and IV were heard and no murmurs. Examination of the respiratory system showed: RR: 32c/m. Cluster breathing pattern, central trachea and vesicular breath sound. Examination of other systems was normal.

Brain imaging (figure 1) showed marked posterior circulation insufficiency with significant atrophy of both occipital lobes as well as the cerebellar structures. Angiography showed numerous abnormal hazy looking vessels arising from the level of the circle of willis and equivalent non filling in the posterior supratentorial and infratentorial spaces typical of moya moya pattern.

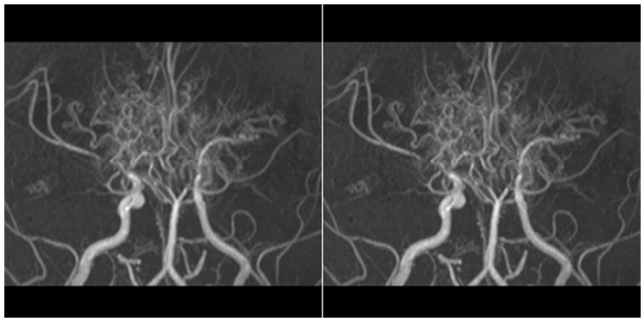

Figure 1. *MRA revealed an abnormal hazy looking vessels arising from the level of the circle of willis and equivalent non filling in the posterior supratentorial and infratentorial spaces typical of moya moya pattern.*

Laboratory test revealed an abnormal HbAIc value of 9.7%, and normal Hb – 14.2g/dl, WBC was 13,400/L. Urine culture and sensitivity: yielded moderate growth of coliform spp. Sensitive to ceftriaxone and streptomycin. Serum electrolyte, urea and creatinine and fasting lipid profile were within the reference range. He was managed as a case of subacute right hemispheric ischaemic stroke secondary to moyamoya disease and septicaemia.

Patient was admitted into the intensive care unit and was nursed as an unconscious patient. Nasogastric tube was also placed for feeding and administration of oral medication. He was commenced on intravenous infusion of normal saline, iv antibiotic, iv antiseizure (levetiracetam), iv omeprazole, oral statin, antihypertensives, haematinics, subcut insulin, subcut clexane, and DVT stocking.

Patient showed clinical improvement with residual neurologic deficit after 7 days of admission in intensive care unit. He was later transferred to the medical ward for continued management. We also commenced discussion with the Family on Prognosis and need to arrange for long term Hospice care. He was later discharged to Hospice care.

3. Discussion

Moyamoya disease (MMD) is a rare cause of ischaemic stroke in our environment. The disease is characterized by a progressive stenosis of the main intracranial arteries. The internal carotid arteries are more commonly affected. This may result in either an ischaemic or haemorrhagic stroke [1]. The disease is seen commonly in Asian populations but has been found in all races with varying clinical presentations. Cho et al. reported an annual stroke rate of 4.5% among 241 hemodynamically stable patients with MMD over 83 months. The annual stroke rate was higher in the hemorrhagic presentation group (5.7%) than the ischemic presentation group (4.2%) or the asymptomatic group (3.4%). They found familial disease as the main risk factor for stroke occurrence [9]. As for ischemic presenting MMD, 5.6% of the annual ischemic stroke rate also showed that posterior circulation involvement was a strong risk factor for ischemic stroke [10]. Our patient had posterior circulation involvement which was responsible for the observed neurologic deficit. Moyamoya disease has been under-recognized as a cause of ischemic strokes in our population due poor access to healthcare and unavailability of sophisticated diagnostic equipment required for early recognition of the disease. The disease is also rare in Africa with reported incidence of strokes in Africa as high as 1-3/100 per year. Abel et al in 2012 reported a case of moyamoya disease in patient with sickle cell anaemia. An association may exist between sickle cell anaemia (as well as other haemoglobinopathies) and moyamoya disease in individuals of African descent [11]. However, these were not the case in the index patient.

Moyamoya can occur at any age and was detected on our patient at an older age but is commonly a disease of children and young adult, with a bi-modal age distribution. The disease had been reported mainly in early childhood and Middle age in various studies. Reports of moyamoya disease in elderly patients with stroke are quite rare in the literatures searched [12]. Late presentation in our patient may be due to the asymptomatic nature and compensatory cerebral blood supply by the collaterals.

The vascular changes in moyamoya disease are usually evident on imaging studies such as CT angiogram and High-Resolution Magnetic Resonance Imaging [7]. The finding most suggestive of Moyamoya on MRI is reduced flow voids in the internal, middle, and anterior cerebral arteries coupled with prominent flow voids through the basal ganglia and thalamus from Moyamoya-associated collateral vessels. These findings are virtually diagnostic of Moyamoya [16]. The index case presented with sudden onset headache, left sided hemiparesis, seizure and loss of consciousness. Further examination revealed signs of cerebrovascular event which were later confirmed with neuroimaging. The findings of moyamoya vessels were mainly confirmed on the index patient with cerebral angiography.

Although, the clinical features found on the patient were consistent with ischaemic stroke and was initially confirmed with non-contrast CT scan, further investigation of the patient with magnetic resonance angiography revealed marked posterior circulation insufficiency with significant atrophy of both occipital lobes as well as the cerebellar structure. There were numerous abnormal hazy looking vessels arising from the level of the circle of willis and equivalent non filling in the posterior supratentorial and infratentorial spaces typical of moya moya pattern. Our patient's angiographic finding was classical of moyamoya disease as the cause of his neurological presentation. High-Resolution MRI not only helps in diagnosis of Moyamoya, but has also been found to be useful in detecting atherosclerotic plaques as well as Moyamoya disease progression [7, 8].

In terms of treatment, the patient was managed medically with acute stroke regimen. He showed some clinical improvement with significant residual motor deficit on the left side of the body. In the acute stage, studies have shown that treatment is the same as for brain infarction or spontaneous intracerebral haemorrhage due to other etiologies such as hypertension and diabetes mellitus. However, there are no effective medical therapies for moyamoya disease [13]. The acute management is mainly symptomatic and directed towards reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures which were present on our patient. Kuroda et al. reported a disease progression rate of approximately 20% over 6 years for patient who received only medical treatment [15]. Surgery remains the best available treatment option because it has been reported to improve cerebral blood flow [13, 14]. The index patient could not benefit from surgical revascularization because he was frail and unfit for the procedure. Houkin et al 2004 also reported that revascularization surgery is a vital option for improvement in cerebral perfusion, and to reduce the risk of a subsequent stroke in both pediatric and adult patients [14]. Once a major cerebrovascular event had taken place as observed on our patient, even with treatment, the patient may be left with permanent loss of function so it is

very important to detect and treat this condition promptly.

4. Conclusion

Although moyamoya disease is a rare progressive vasculopathy involving the intracranial arteries, it should be considered as a differential diagnosis in an African patient presenting with clinical features of stroke. In centers were facilities are available, modern brain imaging techniques (MRI, CTA and MRA) should be used as gold standard test for early diagnosis. Cerebral angiography remains the gold standard in diagnosing the disease. Surgical revascularization remains the best available treatment option. Early identification of the disease would facilitate timely institution of surgical treatment, leading to a better neurological outcome.

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