Case Report

A case of Young Stroke due to Moyamoya Disease

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Abstract

Moyamoya disease is a disease in which certain arteries in the brain are constricted. Blood flow is blocked by the constriction, and also by blood clots (thrombosis). A collateral circulation develops around the blocked vessels to compensate for the blockage, but the collateral vessels are small, weak, and prone to bleeding, aneurysm and thrombosis which may result in TIA, recurrent ischemic or hemorrhagic stroke or seizure. The disease may manifest in pediatric age or young adults. In May 2019 we have diagnosed a young lady with Moyamoya disease who presented with right sided hemiplegia, motor aphasia and dysphagia. She was labeled as hypertensive 6 months prior to this event and used to take anti-hypertensive irregularly and gave past history of occasional headache. Her CT scan and MRI of brain revealed left sided ischemic infarct involving frontotemporoparietal region and cerebral angiogram revealed narrowing of left MCA and non-visualization of distal part. There is extensive fine collaterals (Moyamoya vessels) giving the appearance of puffed smoke. The right ACA and MCA were also narrowed with appearance of early collateral vessels. She was treated with aspirin, PPI, NG feeding, antihypertensive medication, physiotherapy, rehabilitation therapy and other supportive care. His condition gradually improved and discharged on 2.7.19. He was referred to Department of Neurosurgery for cerebral revascularization by STA-MCA (superficial temporal and middle cerebral arteries) bypass surgery after stabilization and MR perfusion study.

Key-words: Moyamoya, Young stroke, Ischemic stroke, Hemorrhagic stroke, Revascularization.

Introduction

Young stroke may be defined as neurological deficit suggestive of stroke in an individual between 18-50 years of age. Young stroke is a major public health problem which comprises 10-15% of all stroke patients. Young stroke has a disproportionately large economic burden by leaving victims disabled before their most productive years. The incidence of young stroke has been increasing worldwide recently due to sedentary lifestyle, change in food habit and increased prevalence of obesity and metabolic syndrome. Various factors and causes are associated with young stroke. Diabetes mellitus, dyslipidemia, smoking, hypertension, obesity, metabolic syndrome, sedentary lifestyle, genetic and familial factors are responsible for majority of strokes in 30s and 40s. Other factors in 20s and 30s and older age groups include valvular heart disease, congenital heart disease like patent foramen ovale, infective endocarditis, cardiomyopathy, atrial fibrillation, prothrombotic conditions like antiphospholipid syndrome, protein C, protein S, antithrombin III deficiency, factor V Leiden, hematological malignancy, polycythemia rubra vera, sickle cell disease, SLE, coagulopathy, bleeding disorders, primary vasculitides, cerebral AV malformation, dissection of carotids, isolated cerebral vasculitis, illicit drugs like cocaine and amphetamine, anticoagulant and blood thinners, epilepsy, migraine, pregnancy, postpartum period, eclampsia, Moyamoya disease etc[1]. Stroke due to Moyamoya disease in particular, may occur at any age ranging from 6 months to 67 years, with the highest peak in the first decade and smaller peaks in the third and fourth decades of life. We have diagnosed a 38-year-old lady with ischemic stroke due to Moyamoya Disease in the Dept. of Neurology, CMH Dhaka on June 2019. This case report has been published to generate awareness regarding the Moyamoya disease and other rare causes of stroke in young.

Case Report

Mrs. X, 38-year-old female housewife, hypertensive for 6 months, non-diabetic, non-smoker, non-alcoholic, hailing from Lakshmipur, was admitted in CMH Dhaka on 24 May 2019 with right sided hemiplegia, motor aphasia and dysphagia for 4 days. On 20 May 2019, she woke up at midnight and fell on the floor while trying to get up from bed. Since then she developed the above-mentioned symptoms. She had history of intermittent headache but not associated with morning wake up or lying down or photophobia/ phonophobia. There was no history of fever, vomiting, convulsion, loss of consciousness, blurring of vision, abnormal movements, chest pain, dyspnea, palpitation, head injury or weight loss. The patient was labelled as hypertensive only 6 months prior to this admission and she was taking Tab Bisoprolol and Tab Losartan irregularly since then. She also reported of taking subcutaneous injectable contraceptives for once about 10 years back.

Upon admission, an extensive physical examination was done with no significant finding on general examination. Carotid or renal bruit was not heard. On nervous system examination, we found motor aphasia, right sided upper motor neuron type of facial palsy, right sided hemiplegia (muscle power 0/5) with hemisensory loss, impaired co-ordination and plantar extensor on right side. All other modalities of nervous system examination and other systems were normal including a normal fundoscopy. With these presentations, the patient was initially diagnosed as Acute Stroke with Right sided hemiplegia with motor aphasia with Right sided 7th nerve

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palsy (UMN type) with HTN. Keeping in mind the wide array of causes of young stroke, several investigations were done. Routine investigations like CBC, RBS, lipid profile, ECG, Urine R/M/E, S. creatinine and electrolytes, chest X-ray were all within normal limits. Other investigations like LFT, S. urea, echocardiogram, Thyroid function tests and coagulation profile were also normal. She was also screened for possible causes of vasculitis, syphilis and other blood dyscrasias with tests such as ANA, anti-phospholipid antibody, p-ANCA and c-ANCA, VDRL, TPHA, protein C, protein S and anti-thrombin III, all of which came negative.

Imaging tests showed significant positive findings. CT scan of brain (Figure-1) showed faintly hypodense area in left fronto-parietal and temporal lobes suggestive of left sided acute cerebral infarct. To get a more vivid picture, MRI of brain (Figure-2) was done which revealed signal intensity change area in fronto-temporo-parietal lobes involving cortical & white matter regions including cingulate gyrus & capsulo-ganglionic region of left cerebral hemisphere. It appeared hypointense on T1W, hyperintense on T2W and FLAIR image- at left ICA territory. Further imaging was done with MRA of brain (Figure-3) which showed non-visualization of Left ICA. Duplex study of neck vessels and brain showed no visible thrombus in major arteries and normal caliber and blood flow in left ICA. However, transcranial colour-coded duplex sonography showed severely blunted flow in left MCA and severe stenosis in right MCA. Cerebral angiogram (Figure-4) was done eventually which showed diffuse narrowing of proximal left MCA & non-visualization of the distal part. There were extensive fine collaterals giving the appearance of puffed smoke. Narrowing of the caliber of right ACA & MCA with appearance of early collateral vessels and narrowing of PCA was also seen.

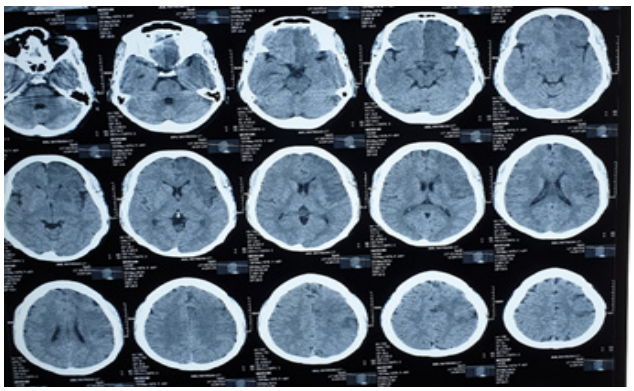

Figure-1: CT scan of brain showing left sided acute cerebral infarct

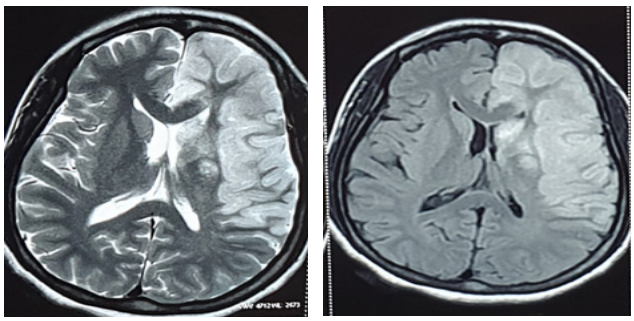

Figure-2: MRI of brain showing left sided acute cerebral infarct

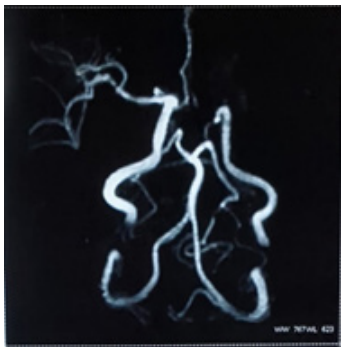

Figure-3: MRA of brain showing non-visualization of left ICA

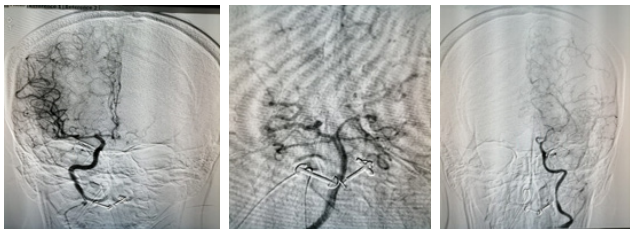

Figure-4: Cerebral angiogram showing the extensive fine collaterals with puffed smoke appearance

Therefore, she was finally diagnosed as a case of Moya Moya Disease with Acute Ischaemic Stroke with RSH with HTN. She was given NG feeding 2 hourly, total 10 feeds daily and was advised regular active physiotherapy, change of posture 2 hourly and care of bowel, bladder and oral cavity. She was started on Tab Ecosprin 75 mg one tablet daily, Tab Losartan 50 mg daily at night. She was referred to Neurosurgery department for Cerebral revascularization by left STA-MCA (EC–IC) bypass surgery after MR perfusion study. The patient was eventually discharged on 2 July 2019 with the above-mentioned treatment. Patient had already improved significantly prior to discharge to a state that she could swallow both liquid and semisolid food and could move the fingers of her right hand. She returned for OPD follow-up on 22 July 2019 where we found her muscle power to have improved to 2/5 on right upper limbs and 3/5 on right lower limbs. She could also walk a few steps with assistance from others.

Discussion

Moyamoya is characterized angiographically by the progressive non-atherosclerotic stenosis or occlusion of the terminal ICAs with the development of collateral networks, which seemingly provides an alternative route for cerebral perfusion. The name “Moyamoya” means “puff of smoke” in Japanese, a term describing the appearance of this cluster of tiny blood vessels. This pathophysiological bypass is imperfect. The attempt to compensate for reduced cerebral perfusion often fails, leading to the well-known clinical manifestations of the disease, including ischemic strokes and TIAs. Moreover, the fragile collateral networks have a propensity to bleed, which may result in hemorrhagic strokes[2]. When Moyamoya is diagnosed by itself, with no underlying correlational conditions, it is diagnosed as Moyamoya disease. This is also the case when the arterial constriction and collateral circulation are bilateral. Moyamoya syndrome is unilateral arterial constriction, or occurs when one

of the several specified conditions is also present[3]. This may also be considered as Moyamoya being secondary to the primary condition.

Moyamoya disease occurs primarily in Asians but can also occur (with varying degrees of severity) in whites, blacks, Haitians, and Hispanics. The female-to-male ratio of moyamoya disease is 1.8:1. Ages for patients with moyamoya disease range from 6 months to 67 years, with the highest peak in the first decade and smaller peaks in the third and fourth decades[4]. The cause of Moyamoya disease is not known. The disease is believed to be genetic. Fukui reported a family history in 10% of patients with the disorder. Moreover, Mineharu suggested that familial moyamoya disease is autosomal dominant with incomplete penetrance that depends on age and genomic imprinting factors[5]. Genetically, susceptibility loci have been found on 3p, 6p, 17q, and band 8q23. People with moyamoya disease have been found to have a higher incidence of elevated thyroid antibodies[6]. It is associated with immunologic diseases like Graves disease and infections like leptospirosis and tuberculosis. Hematologic disorders (e.g. aplastic anaemia, Fanconi anaemia, sickle cell anaemia, lupus anticoagulant etc.) and vascular diseases (Atherosclerotic disease, Coarctation of the aorta and Fibromuscular dysplasia, Cranial trauma, Radiation injury, Parasellar tumors, Hypertension etc.) were also seen associated with Moyamoya diseases. Some congenital syndromes like Alport syndrome, Down syndrome, Marfan syndrome, Tuberous sclerosis, Turner syndrome, von Recklinghausen disease (Neurofibromatosis Type1) and Hirschsprung disease were reported in some cases[7].

Children and adults with moyamoya disease may have different clinical presentations. The symptoms and clinical course vary widely, with the disease ranging from being asymptomatic to manifesting as transient events to cause severe neurological deficits. Adults experience hemorrhage more commonly; cerebral ischemic events are more common in children[8]. Children may have hemiparesis, monoparesis, sensory impairment, involuntary movements, headaches, dizziness or seizures. Mental retardation or persistent neurological deficits may be present. Adults may have symptoms and signs similar to those in children, but intraventricular,

A wide array of investigations has to be kept in mind while evaluating this condition. Coagulation profile must be done to exclude hypercoagulability state – tests include Protein C, Protein S, Antithrombin III, Homocysteine, Factor V Leiden etc. Vasculitis should be excluded by tests like p-ANCA, c-ANCA and autoimmune diseases must be excluded[10]. Cerebral angiography is the criterion standard for the diagnosis of moyamoya disease. The following findings support the diagnosis[11-13]:

1) Stenosis or occlusion at the terminal portion of the internal carotid artery or the proximal portion of the anterior or middle cerebral arteries. 2) Abnormal vascular networks in the vicinity of the occlusive or stenotic areas. 3) Bilaterality of the described findings (although some patients may present with unilateral involvement and then progress). 4) Magnetic resonance angiography (MRA) or computed tomography angiography (CTA) can be performed.

Based on angiography findings, Moyamoya disease can be classified (Figure-5)as following: 1) Narrowing of the terminal ICA. 2) Dilation of the proximal portions of the ACA and MCA with initial basal moyamoya blush. 3) Intensification of Moyamoya vessels, defection of ACA & MCA is observed. 4) Minimization of the basal moyamoya network together with progressive occlusion of the ICA, which reaches the origin of the PCA. 5) Further reduction of moyamoya vessels, with complete disappearance of the main arteries arising from the CA. 6) Disappearance of the moyamoya blush together with the blood supply from the ICA, at this stage, only the ECA supplies the intracranial circulation.

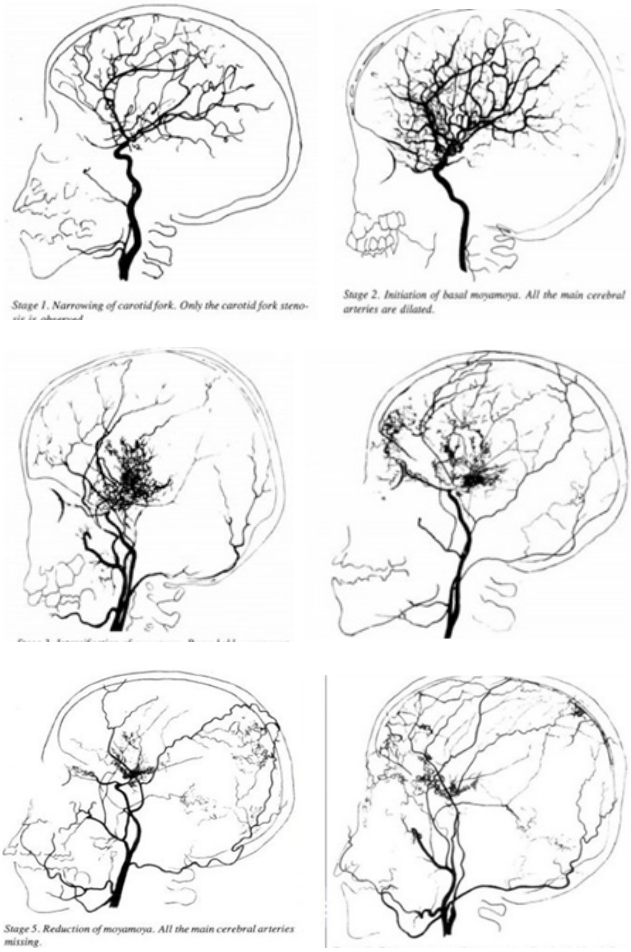

Figure-5: Stages of Moyamoya disease[14].

A characteristic sign for moyamoya on MR imaging is leptomeningeal enhancement. It is related to the leptomeningeal flow engorgement, also known as “ivy sign”. This sign is typically detected with T1-weighted contrast-enhanced and FLAIR images and can be associated with poor visualization of the MCA[15]. The management of Moyamoya disease is largely supportive and symptomatic and depends on the resulting insult. Medical and surgical treatment depending on the insult, along with treatment of associated and comorbid conditions, prevention of complications, physiotherapy and rehabilitation is the mainstay of treatment. The decision to treat patients with anticoagulants such as heparin (and, in some cases, warfarin, for long-term anticoagulation) or antiplatelet agents such as aspirin rests on the following: angiogram findings, severity of stroke, and risk/benefit analysis

by physicians who are experienced in stroke treatment. Surgical treatment is often required. Direct anastomosis by Superficial temporal artery–middle cerebral artery (STA-MCA) anastomosis (EC-IC bypass) is an option. There are also some indirect anastomosis options[16]. Mortality rates from moyamoya disease are approximately 10% in adults and 4.3% in children. About 50-60% of affected individuals experience a gradual deterioration of cognitive function, presumably from recurrent strokes. Patients with moyamoya disease who present for treatment while symptoms are evolving have a better prognosis than do those who present with static symptoms[17].

Conclusion

Moyamoya is a progressive disease that does not have curative treatment. Medical treatment can't prevent the recurrence of stroke, nor fatality once a major stroke or bleeding takes place. Even with treatment the patient may be left with permanent loss of function so it is very important to treat this condition promptly. While Moyamoya itself is not curable, surgery to provide alternative blood flow to the brain prevents the symptoms related to Moyamoya and can provide an excellent long-term outcome with significant stroke risk reduction.

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