Moyamoya disease in a child

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Abstract

A 6 year old boy presented with sign symptoms of cerebral stroke. After clinical and radiological evaluation, a diagnosis of bilateral moyamoya disease was made. An indirect bypass surgery encephaloduroateriosynangiosis was done on the right side at first then on the left side. Patient was partially improved. At one year follow-up, there was the evidence of occlusion of collateral circulation on the right side and the patient became symptomatic again. It is noted that, although the early indirect bypass surgery is indicated in the pediatric patients for preventing the stroke but the outcome is difficult to judge.

Introduction

Moyamoya disease is an occlusive cerebrovascular disorder caused by the blocked bilateral internal carotid arteries at the base of the brain around the basal ganglia.[1,2] It is rare and progressive in nature.[3] Tiny vessels lead to inadequate blood and oxygen supply to the brain. Ultimately oxygen deprivation occurs which causes the signs and symptoms including the seizure, headache, loss of speech, visual disturbances, stroke, paralysis and transient ischemic attack. Moyamoya syndrome, due to similar angiographic appearance, may be linked to the other medical condition such as atherosclerosis, radiation therapy, sickle cell disease, autoimmune disease, brain trauma etc.[5] This disease was firsts reported in Japan in 1957 by Shimizu and colleagues[4] and it is reported that its incidence is higher among the Asian people than in Europe or North America.[1,4]

Because of the rarity of the problem and of clinical interest, we are reporting a case of moyamoya disease

Case Report

A 6 year old boy of non-consanguineous parents presented with a history of suddenly developed blurring of vision accompanied by weakness of the left side of the body with aphasia 2 days back. Weakness was transient and recovered after half an hour. Parents also stated that 3 years back, the boy developed similar type of illness with focal seizure. His learning ability was gradually worsening since then. He had occasional teeth grinding, and urinary incontinence. The boy was immunized and his family history was unremarkable.

On examination, he was afebrile, conscious and mildly anemic. Muscle tone of both the lower limbs were increased. He had no papilloedema and any other neurological deficit. With these complaints parents consulted with the several physicians previously and EEG was also done which showed regional seizure at the left temporal lobe. MRI of the brain was suggestive of multifocal infract in the right centrum-semiovalae and sequeli of vasculities and meningitis (Figure 1).

The boy was treated with antiepileptic drugs but no improvement occurred, rather he developed left sided weakness and was then treated with aspirin. As the boy was not improving and the condition was deteriorating, parents took the decision to go abroad. Several investigations were done including liver function tests, serum lipid profiles, PT, APTT, C3, C4, ANA, antids DNA, plasma ammonia, plasma acilcarnitin, homocystine level, iron profile, urinary amino acid and hemoglobin electrophoresis- all of which were normal. Computed tomography (CT) of brain was done and showed bilateral intraventricular hemorrhage with old infract with the development of confluent arteries in the right white matter and the atrophy of the brain (Figure 2). Diagnostic Magnetic Resonant Angiography (MRA) showed stenosis of supraclinoid segment of both the internal carotid artery with collateral flow in middle cerebral artery (MCA) consistent with moyamoya disease (Figure 3). CT angiography also showed similar results. With these evidences a decision was taken to perform an operation named encephalodurosynaterioangiosis (EDAS) on the

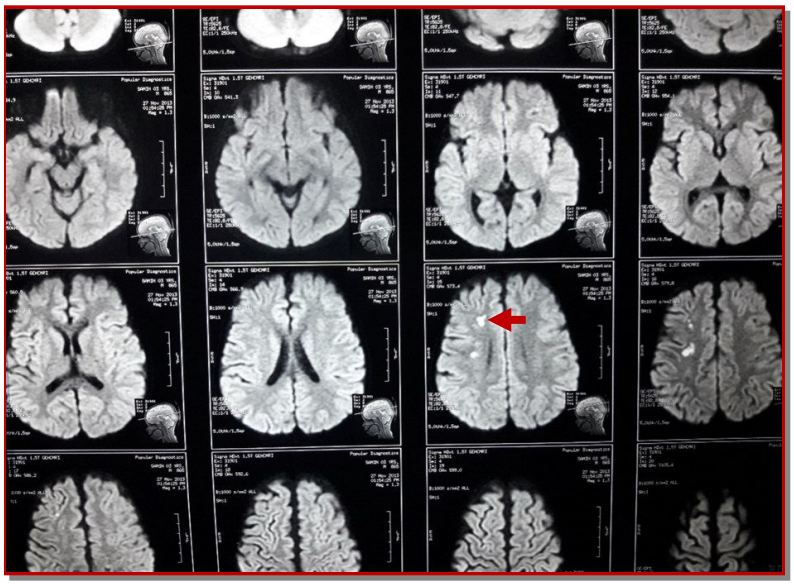

Figure 1: MRI of the brain was suggestive of multifocal infract in right centrum-semiovalae and sequel of vasculities and meningitis

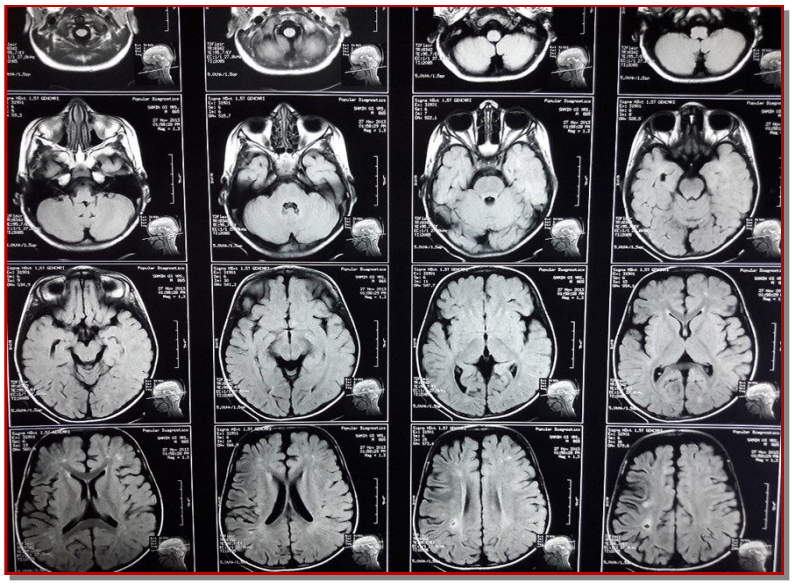

Figure 2: Computed tomography of the brain showed bilateral intraventricular hemorrhage with old infract with development of arterial confluent in right white matter and atophy of the brain

right side at first and then on the left side. After the successful operation, hemiparesis, aphasia and frequency of convulsion were decreased but not totally cured. Patient was advised for regular follow -up. MRI was done after one year showed bilateral pial synangiosis with paucity of the cortical vessels on the right side suggestive of non-functioning anastomosis. Chronic right middle cerebral artery (MCA) territories infract was also noted with decreased blood flow on the right site. After one year, MRI was repeated and showed similar findings. We gave symptomatic management to the patient. Neurosurgery consultation was taken and counseling was done. Patient was discharged with the advice to follow-up.

Discussion

Causes of moyamoya disease is mostly idiopathic, but genetic predisposition seems to be responsible for it, because 1 in 10 individuals with moyamoya disease has a close relative who is also suffered. Most recently 213 (RNF 213) gene in the 17q25 region was identified as the strongest susceptibility gene of MMD in East Asian people.[6] MMD primarily affects children, but it can also affect adults. The average age at which a child is diagnosed is 6 year, but range is 6 month to 8 years.[7] Moyamoya disease in children usually presents with signs and symptoms similar to mini stroke such as– weakness on one side of the body,. slurred speech (dysarthia, aphashia), blurring of vision, cognitive impairment/memory loss, seizure and syncope. Rarely or when the treatment of a child with moyamoya is delayed, the patient may have brain hemorrhage. The warning signs for the hemorrage are: Severe headache, sudden loss of consciousness, vomiting, numbness in part of the body and difficulty in respiration.[7]

In moyamoya disease, MRI not only reveals the areas of infraction, but also visualizes the collateral vessels as multiple blood flow voids at the base of the brain and basal ganglia. Confirmatory angiography shows the narrowing and occlusion of the cerebral vessels and extensive collateral flow, demonstrating the classical puff of smoky appearance.[2, 8, 9] In our case, MRI and MR angiography had the typical 1 findings consistent with moyamoya disease.

In Bangladesh, a few cases on moyamoya in children are reported.[10-16] Some of them presented with hemiplegia and others had occasional seizure. They also had typical radiological findings of moyamoya disease in MR angiogram which consistent with our case.

Medical treatment of moyamoya disease has been utilized to treat many symptoms of moyamoya and is often an important part of the management. But because of obvious risk of cerebral bleeding in moyamoya disease, conservative treatment rarely indicated as long-term measures.[3] Conservatively managed patients with moyamoya disease experienced stroke at a rate ranging from 3.2-15.0%, but after surgical intervention it is usually 0.0-1.6%.[17] So, surgical procedures is preferable choice of treatment and is designed to re-establish blood supply to the brain surface and thereby circumventing the progressive loss of brain hemisphere blood flow.

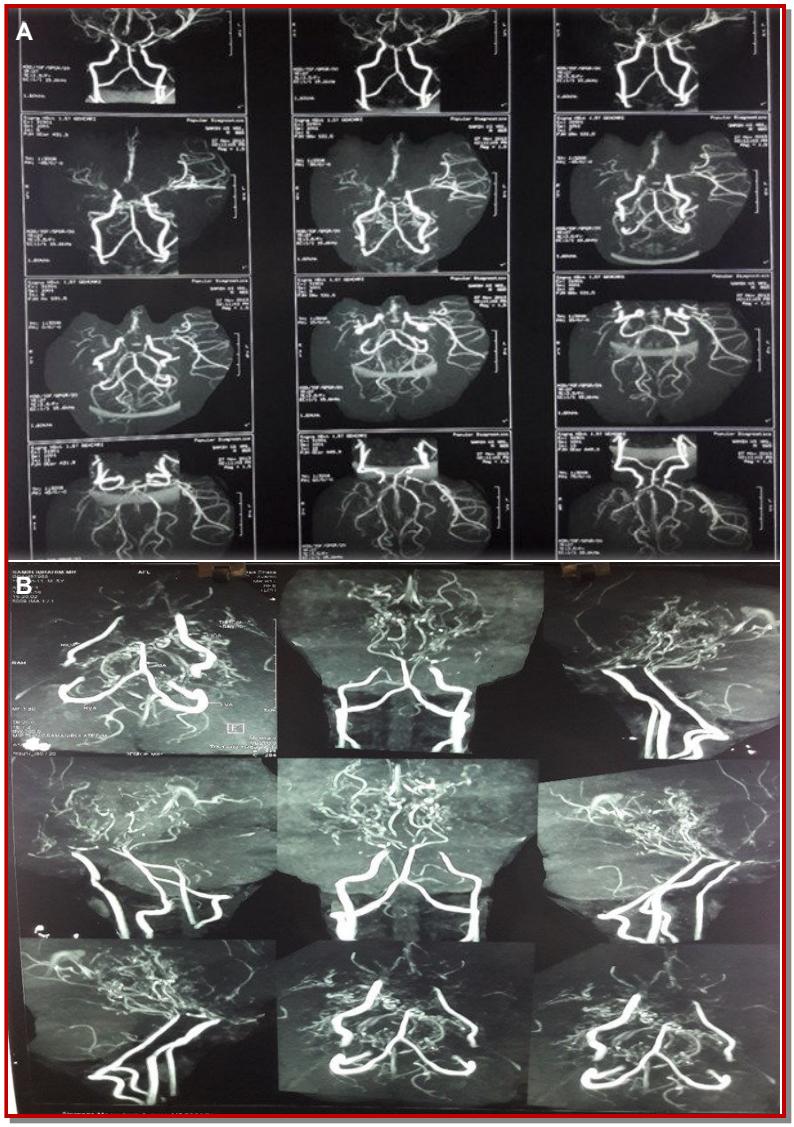

Figure 3: (A+B) Diagnostic Magnetic Resonant Angiography (MRA) showed stenosis of supraclinoid segment of bilateral segment internal carotid artery with collateral flow in middle cerebral artery is consistent with moyamoya disease

There are many surgical procedure proposed to treat the moyamoya. These are indirect and direct procedures. Indirect procedures such as pial syngiosis, encephalomyosyngiosis (EME), encephaloduroarteriosynangiosis (EDAS), dural inversion and other similar variant usually carried out in children and younger patients.[18, 19] Direct revascularization of the pediatric patient with moyamoya disease may also be effective in further stroke.[20] One of the indirect procedures, EDAS was done in our patient. After successful operation although he was improved but symptoms again reappeared due to blockage of collateral circulation. It is reported that the incidence of newly developed cerebral infract after indirect revascularization is somewhat high.[20] In adult cases treated with indirect procedure, up to 14% of patients' experienced postoperative stroke and on the other hand, the pediatric patients showed 1.6% unsatisfactory result.[20, 21] So, indirect procedure for the moyamoya disease is thought to be more effective in the pediatrics patients.

Conclusion

Appropriate treatment for the moyamoya disease is yet to be determined. But due to progressive nature of the disease, surgical treatment for the moyamoya disease should be considered, for symptomatic patients. For the pediatric patient, early diagnosis and surgical intervention is mandatory for preventing control of further stroke and hemorrhage.

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