Case Report

Moyamoya Disease – A Case Report of Six Years Old Girl
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ABSTRACT
A 6 year old girl is with recurrent stroke is described which was diagnosed as Moyamoya disease, emphasizing the need to recognize the condition and treat them early. (Rawal Med J 2008;33:115-117).

Key Words: Moyamoya disease, MRI, MRA.

INTRODUCTION
Moyamoya disease (MMD) is a rare idiopathic disorder that leads to irreversible blockage of the major blood vessels to brain. In Japanese, it means “puff of smoke” which refers to collateral circulation that develops adjacent to the stenotic vessels. The steno-occlusive areas are usually bilateral, but unilateral involvement does not exclude the diagnosis. It can lead severe functional impairment or even death. Familial predisposition is 10% that may be hereditary and multifactorial. The cause of MMD is not known. The disease is believed to be hereditary. Genetically, susceptibility loci have been found on chromosomes 3p, 6p, 17q, and band 8q23. MMD being a rare case in our part of the world should be considered in the differential diagnosis of young stroke. Here we report a case of young girl with Moyamoya disease.

CASE REPORT
A six years old girl developed sudden weakness of right arm and leg at the age of 1½ year that was diagnosed as stroke. She improved slowly and started to speak and walk again. She stopped talking at the age of 4 years following a febrile illness. On examination, she had drooling of saliva, was quiet and looked mentally dull. There was no facial asymmetry; motor system showed that she was using left side more than right, deep tendon reflexes were hyperactive bilaterally and planters were bilaterally flexor. Brain MRI and MRA showed non-visualization of bilateral anterior cerebral artery, middle cerebral artery, supraclinoid part of bilateral internal carotids and multiple collaterals.

**DISCUSSION**

MMD occurs primarily in Asians, but it can occur in Whites, African, Americans, Haitians and Hispanic. The female to male ratio is 1.8:1, age range from 6 months to 67 years with the highest peak in the first decade and smaller peaks in the third and fourth decades. Pathologically, MMD is characterized by intimal thickening in the walls of the terminal portions of the internal carotid vessels bilaterally. The proliferating intima may contain lipid deposits. The anterior, middle, and posterior cerebral arteries that emanate from the circle of Willis may show varying degrees of stenosis or occlusion. This is associated with fibrocellular thickening of the intima, waving of the internal elastic lamina, and thinning of the media. Numerous small vascular channels can be seen around the circle of Willis. These are perforators and anastamotic branches. The pia mater also may have reticular conglomerates of small vessels.

Fig. 1 The patient.
MMD may occur by itself in a previously healthy individual, or rarely be associated with Grave’s disease/thyrotoxicosis, leptospirosis and tuberculosis, aplastic anemia, Fanconi anemia, sickle cell anemia, lupus anticoagulant, Apert syndrome, Down syndrome, Marfan syndrome, tuberous sclerosis, Turner syndrome, atherosclerotic disease, coarctation of aorta, fibromuscular dysplasia, cranial trauma, radiation injury, parasellar tumors, hypertension. Mortality rates are approximately 10% in adults and 4.3% in children.Death is usually from hemorrhage. Adults experience hemorrhage more commonly. Cerebral ischemic events are more common in children. Our patient also had ischemic infarcts of variable ages involving bilateral cerebral hemispheres.

Fig 2. MRA shows collateral circulation through anterior and posterior circulation.
Cerebral angiography is the standard criterion for diagnosis. MRA can be performed and shows collateral circulation through posterior circulation, as seen in our patient. Pharmacologic therapy for MMD is disappointing. Therapy is directed primarily at complications of the disease. If the patient has had an ischemic stroke, anticoagulation or antiplatelet agents should be considered. We treated our patient with antiplatelet agents. Encephaloduroarteriosynangiosis (EDAS) was performed in patients of MMD and no episode of stroke or transient ischemic attack (TIA) were observed in any patient during 2 years follow up period and all patients were living without new neurological deficit. Long term out come of EDAS is promising.

Fig 3. T1 image show wedge shaped areas of encephalomalacia in left mid parietal region, left temporal lobe, posterior frontal region, right posterior parietal and anterior frontal region.
Various surgical procedures including superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis, EDAS, encephaloduroarteriomyosynangiosis (EDAMS), pial synangiosis and omental transplantation have been used. Rehabilitation with physical therapy, occupational therapy and speech therapy should be considered depending on the neurological impairment. The extent of therapy can range from bedside to full comprehensive inpatient care. Vascular surgery input was taken in our patient but no surgical intervention was suggested.

Conclusion

A high degree of suspicion is required to diagnose MMD in children with recurrent stroke. It is important to recognize and treat them early. Cerebral angiography is the most definitive method of diagnosis.

REFERENCES


