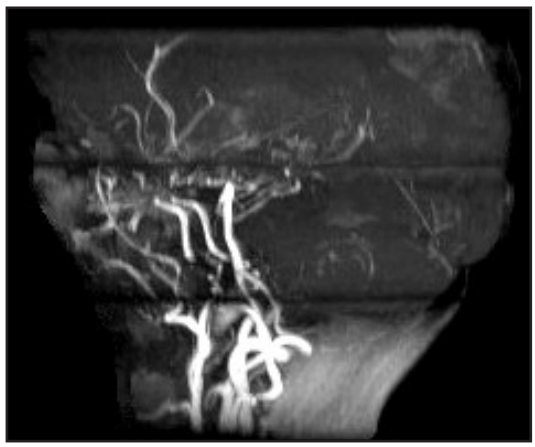
SPOT DIAGNOSIS (IMAGE GALLERY)



Recurrent strokes

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A nine year old boy was presented with sudden onset of inability to move right upper limb, inability to speak and blurring of vision. The weakness progressed to the lower limb one day after the admission. Two days later he started speaking few words accompanied by facial expression and head nodding movements. The child had similar type of sudden onset of weakness of left upper and lower limb at the age of 4 years, but remarkable improvement of weakness occurred over a period of 2 weeks after initiating physiotherapy. During the proceeding 4 years, the child developed repeated attacks of weakness of right upper limb.

On day 3rd of admission, the child became semiconscious, not recognizing even the mother indicating another bout of a stroke and/or extension of the previous attack. Upper motor type right facial palsy also appeared after the fresh attack.

MRI Brain showed acute non-hemorrhagic MCA zone and left super medial fronto parietal ACA zone infarction. Chronic right temporo parietal, occipital, frontal PCA, ACA, MCA, watershed and left frontal watershed infarcts were present.

MRI angiogram is shown in the figure.

What is the diagnosis?

Moya Moya Disease. MRI angiogram shows both the internal carotid arteries reduced in caliber in their intracranial course especially in the cavernous and supraclinoid parts. Multiple thin tortuous collaterals were seen around the Circle of Willis {Puff of smoke sign}.

Moya Moya disease is a rare disorder of the blood vessel of the brain characterized by progressive occlusion of the cerebral vasculature with particular involvement of the Circle of Willis and the feeding arteries. This distinctive name Moya Moya is Japanese for "Puff of Smoke" which describes the angiographic appearance of the abnormal vascular collateral that develops adjacent to the stenotic vessels. Most of the time, the disease affects adults in third to fourth decades of life, but, tends to cause stroke and seizures in children. The clinical features of disease are those of cerebral ischemia {strokes}, recurrent transient ischemic attacks {TIA}, sensorimotor paralysis, convulsions and /or migraine like headache. The reported annual incidence of cerebral infarction in children, all over the world varies between 1.2 to 2.7 per 1,00,000. Though, no definite data is available in India, however, pediatrics stroke constituted less than 1 percent of all pediatric admissions and 5-10 percent of all strokes in young and adults { Less than 40 years}. The constriction of arteries in Moya Moya disease is unlike the constrictions in atherosclerosis. In atherosclerosis, the inner layer of {lumen} the arteries suffer from immune reactions, fills with inflammatory cells, accumulated fatty cells and debris. In Moya Moya disease, the inner layer of the carotid artery overgrows inward to constrict the artery which gets filled with blood clots, leading to strokes. The diagnosis is always suggested by CT, MRI and MRI angiograms. Patients with Down syndrome, neurofibromatosis or sickle cell disease can develop Moya Moya disease. Anti-platelet agents e.g. aspirin are usually given to prevent clot. Surgery is usually recommended in order to direct other arteries such as the external carotid or the superficial temporal arteries to replace the circulation of blood affected by narrowing of internal carotid and other adjacent arteries. Although there is 4 percent risk of stroke soon after surgery, there is 96 percent probability of remaining stroke free over the next 5 years. Though the natural history of this disorder is not well known, the long term outlook for patients with treated Moya Moya seems to be good. Once major strokes or bleeding take place, inspite of treatment, the patient may leave with permanent loss of function. Therefore, it is very important to diagnose and treat this condition promptly.

E-published: July 2010 . **Art**#44