Moyamoya Disease

A 2-yr-old boy presented with recurrent episodes of transient ischemic attacks since the age of 6 months, following which he had delayed milestones. The boy also had history of seizures since one yr. He was referred to MRI of the brain for further evaluation.

Imaging Findings

Fig. (a). T1weighted image at the level of basal ganglia shows multiple, tiny, low intensity rounded areas in bilateral basal ganglia (arrows), which appear hyperintense on T2 weighted image (b) suggestive of collateral vessels (c) Fluid attenuation inversion recovery (FLAIR) sequence showing atrophy involving bilateral frontal (arrows) and left parietal cortical areas consistent with chronic ischemic changes (d) Volume rendered 3D time of flight (TOF) frontal projection cerebral angiography showing significant stenosis in the supraclinoid internal carotid arteries on both sides with absent flow in the anterior and middle cerebral arteries and multiple collaterals in bilateral basal ganglia appearing as "puff of smoke" (moyamoya) (arrowheads). Cause is unknown in this boy.

DISCUSSION

The term 'moyamoya' means 'wavering puff of smoke', described by Takaku, Suzuki and others in Japan in the sixties.¹ Moyamoya syndrome is a phenomenon caused by an oligemic state occurring in various disease entities.

Chromosomal disorders (Neurofibromatosis, Down's syndrome), hemolytic anemias, infectious diseases (leptospirosis, tuberculous meningitis), neoplasms (Craniopharyngioma, Wilm's tumor), drug abuse (phenobarbitol). There have been recent reports of increased familial incidence of the disease with markers on chromosome 6 and 17, located at 3p24.2-26 and a possible linkage of the marker D6S441 to the disease on chromosome 6. Moyamoya is commonly seen during first decade of life. Frequently, the children present with transient ischemic attacks or ischemic strokes. Cerebral ischemia may also present as reversible ischemic neurological deficit, sensory attacks or acute infantile hemiplegia. Headaches, involuntary choreiform movements, seizures and other motor disturbances have been described.²

MRI and MR angiography are diagnostic imaging modality of choice. MRI shows multiple dot-like flow voids in basal ganglia on T1 weighted image (WI). Flow voids in the multiple collateral vessels in the cisterns appearing as 'net like' cisternal filling defects, associated with hyperintense small-vessel cortical and white matter infarcts which are seen on T2WI. Slow flowing engorged pial vessels with thickened arachnoid membranes appear as bright sulci called leptomeningeal "ivy sign". MR angiography shows narrowed distal ICA and proximal Circle of Willis vessels with multiple lenticulostriate and thalamoperforator collaterals giving appearance of a "puff of smoke". MR spectroscopy will show lactate peak in acutely infarcted tissue. Treatment is mainly Encephaloduro-arterio-synangiosis (EDAS), which is a method of indirect bypass.³

Teaching point : Moyamoya disease or syndrome should be suspected in a child with recurrent ischemic attacks. MRI with MR angiography is the investigation of choice in this condition.

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