Moyamoya Disease with Hypertension in a Young Adult
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Abstract
A 13 year old boy presented with recurrent TIAs and was found to have high blood pressure. On investigations he was detected to be Moyamoya disease. Hypertension is known to be associated with Moyamoya disease; the cause being renal artery stenosis. Our patient had normal renal arteries on imaging. We report this case for its rarity.

Introduction
Moyamoya disease is the progressive vasculocclusive disease of unknown cause involving arteries of the ‘circle of Willis’. It presents as ischemic stroke in children and intracranial hemorrhage in adults. It is associated with renal artery stenosis and renovascular hypertension in some patients.

Case Report
A thirteen year old boy, 8th standard student, presented with sudden onset weakness of left upper and lower limb one month back. The weakness had improved spontaneously in 15 minutes. He was found to have high blood pressure and was started on cap. Nifedipine by a general practitioner. In one month he had four such episodes of left sided transient weakness followed by complete recovery. On enquiry he also had two episodes of transient blackout in right eye 2 months ago with complete recovery.

There was no history of convulsion or unconsciousness or trauma. There was no history of paroxysms suggestive of pheochromocytoma. There was no history of oliguria or edema.

On examination pulse was 86/min, regular, all peripheral pulses including carotids were normal. Blood pressure was 170/100 mm of Hg. There was no pallor, edema feet, skin rash, arthritis, oral ulcers. Cardiovascular and respiratory system examination were normal. No renal bruit was found per abdomen. Neurologically higher mental functions were normal. Cranial nerves were normal. Fundoscopy revealed grade I hypertensive retinopathy. Left side hemiparesis was present with muscle power being grade 3/5. Sensory examination was normal. CT scan of brain showed acute infarct in territory of right middle cerebral artery. The MRI brain showed subacute infarct in right frontotemporal and subcortical white matter, old infarcts in right parieto-occipital region and old hemorrhagic areas in bilateral basal ganglia.

MR angiography showed multiple focal narrowing involving right internal carotid artery with absence of flow signal in right middle cerebral artery. The right anterior cerebral artery filled up by retrograde filling. A1 segment of right ACA, M1 segment of right MCA and P1 segment of right and left PCA showed focal narrowing (Fig. 1). These findings were suggestive of Moyamoya disease. Digital subtraction angiography of cerebral arteries showed occlusion of supraclinoid internal carotid artery, normal vertebro-basilar circulation, posterior communicating artery supplying middle cerebral artery, anterior cerebral artery formed by pial collaterals and normal left side arteries (Fig. 2). The impression was chronic progressive occlusive arteriopathy suggestive of Moyamoya disease.

He was started on antiplatelet medication, Atorvastatin and B.P. was controlled with Amloidepine, Atenolol and Prazosin. The investigations were done for secondary causes of hypertension as patient was young. The reports were as follows: Hb-16g/dl, WBC count-8800/cumm, platelet count-3,63,000/cumm, PS-normocytic, normochromic, ESR-25, Sr.Creatinine-0.9mg/dl, BUN-11mg/dl, RBS-100, Na-139mEq/l, K-3.8mEq/l, X-Ray chest normal, ECG-normal, Echocardiography-normal, Liver function tests-normal, Urine-albumin trace. 24 hour urine proteins were absent. USG abdomen-normal, Renal artery Doppler-normal. There was no evidence of luminal or osteal narrowing or dilatation of renal arteries. Renal angiography done subsequently was normal. 24 hour urine metanephrine-85mcg (normal:25-312), Serum metanephrine-normal, Sr.cortisol-normal, TSH-normal, Sr. Homocysteine-1.02mcg/ml(normal). ANA, anti ds-DNA, ANCA were negative, APLA-negative (IgG-1.54GFLU/ml, IgM-1.66MFLU/ml), C3-155mg/dl (88-201mg/dl), C4-27mg/dl (16-47mg/dl). Protein C, Protein S, Antithrombin III-negative. Sickling test-negative. Lipid profile-total cholesterol-246mg/dl, HDL-48mg/dl, LDL-157mg/dl, VLDL-40mg/dl, Triglyceride-200mg/dl. CT abdomen did not show any adrenal humour.

Presently patient’s BP is controlled with medications; residual weakness is present in left upper and lower limb. Patient and his parents are not willing for neurosurgical procedure.

Discussion
Moyamoya disease is a progressive occlusive disease of cerebral arteries particularly involving the ‘Circle of Willis’ and the arteries feeding it. The network of vascular collaterals developing adjacent to stenotic vessels gives rise to the appearance of ‘puff of smoke’ (Moyamoya in Japanese) on angiography. The stenoses are usually bilateral. But unilateral involvement does not exclude the disease.

The incidence of Moyamoya disease is the highest in Japan; 0.54 new cases per one lac population per year. The prevalence is 3.16 per lac population. Females are affected more commonly than males. Female:Male ratio is 1:1.1. The age of presentation shows bimodal peak i.e.-in first decade and between 3rd and 4th decades.

The etiology is unknown. Genetic backgrounds have been suggested in familial occurrence. The responsible genetic locus have been located on chromosomes 3,16,17. Angiogenic cytokines may play a role in the progressive steno-occlusive changes and/or in angiogenesis of collaterals. The characteristic histopathological features of the stenosed arteries are fibrocellular thickening of intima containing proliferated smooth cells and prominently tortuous and often duplicated internal elastic lamina. There is usually no atheromatous plaque in the arterial wall.

Moyamoya disease is associated with following diseases: 1.
1. Immunological: Grave’s disease, Thyrotoxicosis.
2. Infections: Leptospriosis, Tuberculosis.

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Received: 19.03.2009; Accepted: 01.12.2009
Fig. 1: MR angiography showing multiple focal narrowing involving right internal carotid artery with absence of flow signal in right middle cerebral artery, right anterior cerebral artery filling up by retrograde filling, focal narrowing of A1 segment of right ACA, M1 segment of right MCA and P1 segment of right and left PCA, normal vertebral and basilar arteries.

5. Vascular: Atherosclerosis, Coarctation of aorta, Fibromuscular dysplasia, cranial trauma or irradiation, parasellar tumours, Hypertension.

These associations are not necessarily causative.

Clinically the symptoms of cerebral ischemia dominate in children and intracranial hemorrhage is more common in adults. The ischemic attacks in children are often provoked by hyperventilation. Recurrent strokes lead to permanent neurological deficits and may cause gradual deterioration of cognitive function. Moyamoya disease is the cause of cerebrovascular disease in 5.9% of cases in children though it is less common in adults. The unilateral Moyamoya disease has lower frequency of strokes as compared to bilateral (typical) disease.

Cerebral angiography is the gold standard for diagnosis of Moyamoya disease. The findings are:
- Stenosis or occlusion of terminal portion of ICA or proximal portion of ACA or MCA.
- Vascular collateral network in vicinity of stenosed areas giving rise to ‘puff of smoke’ appearance.
- Bilaterality of these findings. Some patients may present with unilateral findings and then progress.

Nowadays diagnosis is also done on MR angiography.

Medical therapy is directed at complications of the disease. BF is controlled with antihypertensives in case of intracerebral hemorrhage. In case of ischemic stroke antiplatelet agents and anticoagulants are given to prevent future strokes. Since there is risk of hemorrhage from fragile collateral vessels the decision regarding use of anticoagulants depends upon the severity of stroke, angiographic findings and risk-benefit analysis by experienced physician. Various anastomotic surgical procedures are useful to prevent ischemic stroke. Their usefulness to prevent hemorrhage is not known.

Hypertension is associated with Moyamoya disease in 5% of patients. In most of such cases the cause of hypertension is found as renal artery stenosis. In two Japanese studies the incidence of renal artery stenosis in patients with Moyamoya disease was 5% and 7%.

Our patient did not have renal artery stenosis on Doppler as well as on angiography. We report this case for its rarity.

References
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