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CLINICAL BRIEF

Moyamoya Disease with Renal Artery and External Iliac Artery Stenosis

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Abstract Moyamoya disease is a rare, progressive occlusive disease of the cerebral vasculature, mainly involving internal carotid and proximal cerebral arteries with development of fine collateral vascular network in brain. Coexistence of renal vascular lesion with cerebral vascular lesion has rarely described and association with external iliac and femoral vascular stenosis is not known to the best of our knowledge. This is the first case of renovascular hypertension with Moyamoya disease being reported in India with involvement of other extra cranial vessels.

Keywords Moyamoya disease · Renal artery stenosis · Renovascular hypertension · External iliac and femoral artery stenosis

Abbreviations

MMD	Moyamoya disease
RVHT	Renovascular hypertension
RAS	Renal artery stenosis
ICA	Internal carotid artery
ECA	External carotid artery
ACA	Anterior cerebral artery
PCA	Posterior cerebral artery
MCA	Middle cerebral artery

Introduction

Moyamoya disease is a rare progressive occlusive disease of cerebral vasculature, mainly involving internal carotid (ICA) and proximal cerebral arteries with development of fine collateral vascular network in brain giving a "puff of smoke" appearance on Angiography (Japanese Moyamoya). Extracranial vessel involvement especially renal artery is known but is extremely rare. This is the first published case of renal artery stenosis with Moyamoya disease from India with involvement of external iliac and femoral vessels.

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Case Report

An 8 yr-old-child, known case of Moyamoya disease presented for evaluation of uncontrolled hypertension. At 3.5 years of age he developed acute onset left sided focal seizure followed by left sided hemiplegia and ipsilateral upper motor neuron facial nerve palsy. There was no history of any developmental delay or seizure disorder in family and he was normotensive. CNS examination revealed left sided hemiplegia with UMN facial nerve palsy with normal other systemic examination. His thrombophilia workup was normal. MR Angiography (Fig. 1) confirmed vascular etiology of disease and showed narrowing of both internal carotid artery (Lt>Rt) extending up to external carotid and middle carotid vessels. Diffuse irregular narrowing in Rt. Intra and extracranial ICA, B/L proximal Middle cerebral artery (MCA), A1 segment of Anterior cerebral artery(ACA) with multiple collateral between Rt. ECA and ICA are typical of Moyamoya disease (lt>rt). It also showed involvement of ECA, mainly superficial temporal and lt IMA and also collateral from Posterior cerebral artery.

Parents were counseled for shunt surgery but parents did not consent. He received regular physiotherapy and showed gradual improvement with clinically minimal neurological deficit after 1 yr. On follow up a year later,

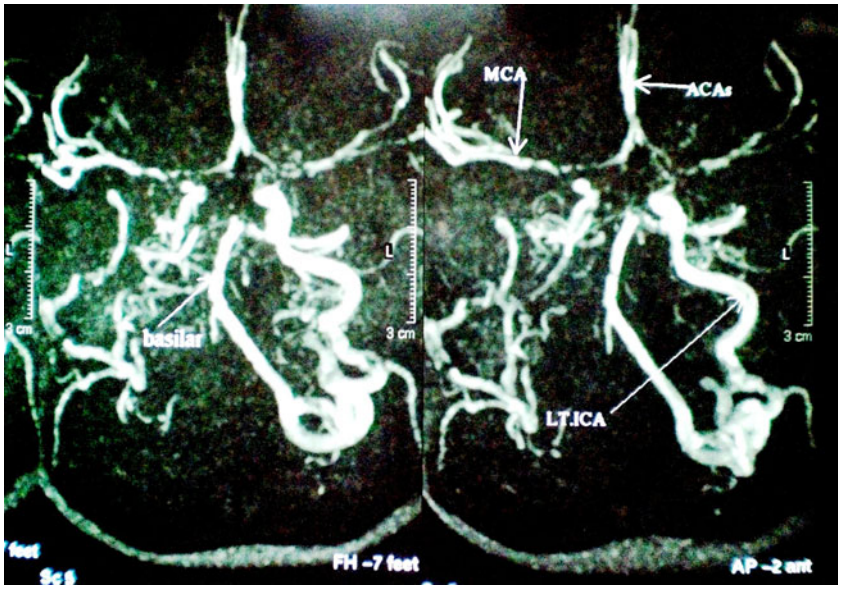

Fig. 1 CT Scan at 3–5 year showing stenosis of left ACA, MCA and Moyamoya collaterals

hypertension (BP 134/90 mmHg) was noted with no neurological worsening of disease. Routine BUN and creatinine were normal (11 and 0.38 mg/dl, respectively). Renal color Doppler showed significant stenosis of left renal artery at mid portion. This finding was later confirmed on MR angiography of abdominal vessels. (Fig. 2) which in addition showed, narrowing/occlusion of lt External iliac and lt Common femoral vessels but Aorta contour and other abdominal branches were normal with normal renal sizes.

Child was put on nefidepin (2 mg/kg/day) but lost to follow up. At 8 yrs of age he presented with a limp. There were stigma of old hemiplegia and limb length discrepancy of 3.8 cm. His blood pressure in upper limb was 160/100 mmHg(non compliance and improper dose of nefidepine for present weight). Left lower limb pulses like dorsalis pedis and posterior tibial were present but weak as compared to right side but femoral artery could not be palpated. Repeat color Doppler showed the same lesion with no progression of stenosis. Renal functions remain normal with creatinine 0.6 mg%. Captopril DTPA angiogram did not show hemodynamically significant obstruction. Blood pressure now controlled with proper dose (2 mg/kg of nefidepine) without any interventional angioplasty.

Discussion

Moyamoya disease is largely believed to be intracranial vacuities of unknown etiology involving the arteries of circle of Willis, with abnormal capillary network formation resulting in ischemic or hemorrhagic stroke.

The disease is usually sporadic, however familial cases (up to 7% of total cases) are also known especially in Japanese population with linkage to region 3p24–26 [1]. An acquired form of disease has also been described, sometimes in association with sickle cell disease, Down's syndrome, nephrotic syndrome [2].

Pathologically, blood vessels show intimal fibromuscular dysplasia and lipid deposition and infiltration with inflammatory cells. Those changes are seen not only in carotid and cerebral vessels but also involve coronary, renal arteries, pulmonary and pancreatic arteries too, leading to the theory that it may actually be an intracranial manifestation of systemic arterial disorder [3].

Co-existence of cerebral and renal lesion is rarely reported. Yamada et al. prospectively examined 86 patients of idiopathic Moyamoya disease with both cerebral and abdominal angiography and found 8% were having renal artery lesion, 7% had stenosis in renal artery and 1% had a small secular aneurysm [4]. Cerebral aneurysm is seen mostly in adult form of disease but it is extremely rare in renal vessel (only 2 cases describe so far).Renal aneurysm may be associated with cerebral aneurysm [4]. Most of the patients develop hypertension later (as in our patient) but 2 who had marked renal artery stenosis and presented with renovascular hypertension. There was no significant correlation between renal artery and cerebral angiography lesion. A similar study by Osamu et al. [5] reported an incidence of 5% of renal artery lesion (out of 73 patient), with 3% having moderate stenosis and renovascular hypertension.

Renal artery stenosis may be progressive and hypertension may develop later, hence a close follow up is required. The hypertension in our patient was easily controlled on

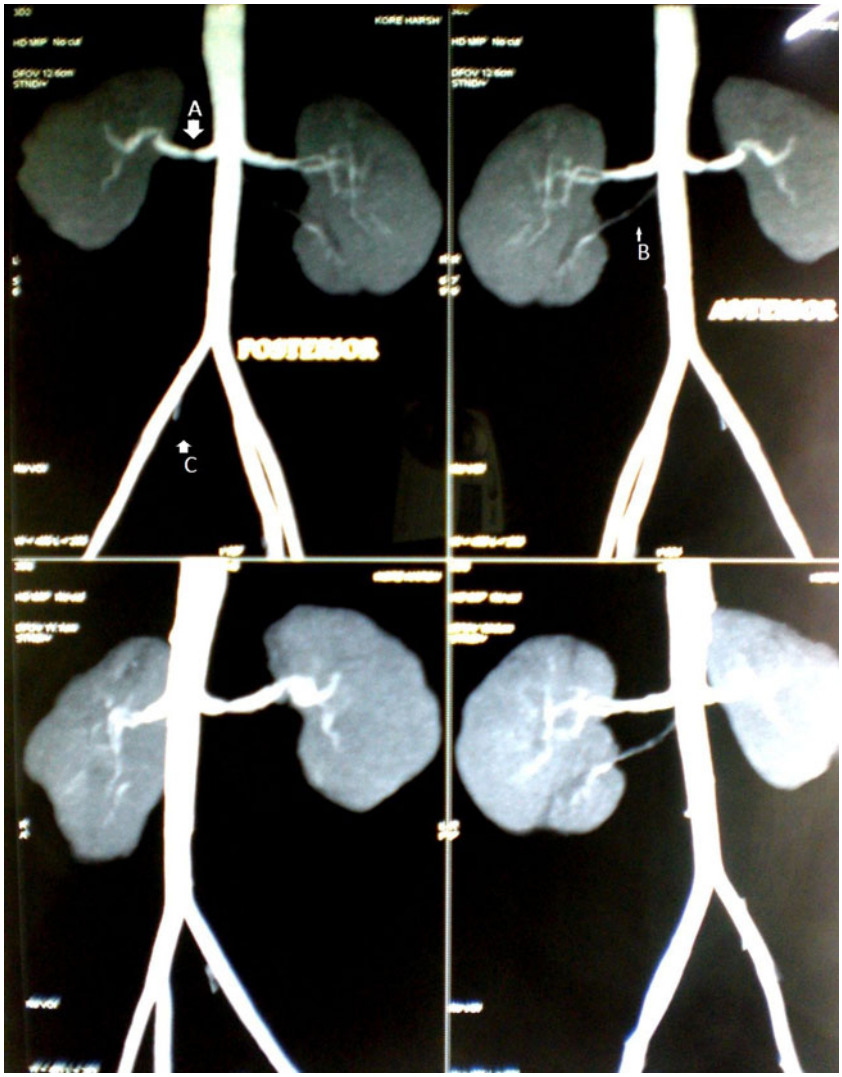

Fig. 2 MR angiogram showing narrowing of lt renal artery at origin (**A**) also multiple narrowing of both renal vessels including Rt. accessory renal artery (**B**) in mid and distal segment. There is also narrowing/complete occlusion of lt. external iliac and common femoral artery (**C**)

minimal antihypertensive and captopril scan also did not show hemodynamically significant obstruction either, hence we are only following the child and no Interventional procedure has been done. Renal angioplasty as a intervention procedure for controlling hypertension has been described. Involvement of external carotid artery is very rare in Moyamoya disease [6].our patient has involvement of not only External carotid artery but also its branches like superficial temporal artery. External carotid system involvement has clinical importance, that it may lead to failure of ECA and ICA shunt [6]. Our patient has narrowing/occlusion of lt External iliac and lt Common femoral artery which has never been describe in association with Moyamoya disease. Although it was asymptomatic without any intermittent claudication, it may have contributed to already decreased growth of hemiplegic limb causing severe leg length discrepancy.

Treatment

Shunt surgeries with aim to provide the blood flow to the ischemic area, have favorable result in preventing progression of CNS symptoms [7] especially motor stroke and TIA. Interestingly conservative management without surgery in adult has also been described with at least 40% not bleeding in mean follow up of 12 yrs, but 60% having re-bleeding and worse outcome with highest risk been age >36 yrs [8]. Renal artery stenosis can be very well treated with angioplasty in most of the cases [4], sometimes requiring auto transplantation and rarely requiring nephrectomy [9].There is debate whether to operate brain lesion first or control BP [10], as disturbed cerebral hemodynamic reserve in these patients, use of PTA or antihypertensive before treatment of brain lesion, may cause precipitous fall in BP leading to cerebral ischemia.

Contributions Dr. Javed Ahmed was responsible for literature search, manuscript writing, and management of the patient; Dr. Uma S. Ali was responsible for diagnosis, supervising and planning the management of patient.

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