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Case report

Renal agenesis and external iliac artery stenosis in an infant with moyamoya disease

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Abstract

We describe a 14-month-old girl who presented with arterial ischemic stroke due to moyamoya disease, unilateral renal agenesis and external iliac artery stenosis. The association of moyamoya disease with renal agenesis and external iliac artery stenosis has not been described before. This report expands the spectrum of moyamoya disease and suggests that moyamoya disease may have an intrauterine onset.

Keywords: Stroke; Childhood; Circle of Willis; Vasculopathy

1. Introduction

Moyamoya disease is a rare cerebral vasculopathy of unknown etiology, characterized by stenosis or occlusion of the supraclinoid portion of internal carotid artery and a prominent collateral network at the base of the brain (seen as "puff of smoke" on angiography) [1]. Renal artery is the commonest extracranial vessel affected, involved in 5–8% of patients [2,3]. There are very few reports of involvement of other extracranial arteries [4–8]. We report a young child who had moyamoya disease along with unilateral renal agenesis and external iliac artery stenosis.

2. Case report

A 14-month-old girl was born at term by caesarean section in view of maternal oligohydramnios. Her developmental milestones have been achieved at appropriate ages. She suffered sudden onset right hemiparesis, flurry of seizures and altered sensorium. She was admitted at a hospital where she received phenobarbitone, phenytoin and midazolam infusion. Two days later, she developed left hemiparesis. She was also detected to have hypertension, which was treated with sodium nitroprusside and amlodipine. She was subsequently referred to our hospital. Family history was unremarkable.

Examination at admission revealed heart rate of 160/min and blood pressure of 120/78 mm Hg (>99th centile). Neurological examination revealed spasticity in all limbs (right side more spastic than left), and reduced muscle power with 3/5 and 2/5 on the right and left sides, respectively. The deep tendon reflexes were brisk with bilateral extensor plantar response.

Ultrasonography and renal dynamic scan using LL-ethylene cysteine showed normally functioning left kidney and non-visualized right kidney. Brain magnetic resonance imaging (MRI) revealed diffuse cerebral atrophy, left middle cerebral artery territory infarct and attenuated flow voids of internal cerebral and middle cerebral arteries (Fig. 1A and B). Digital subtraction

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angiography (DSA) showed irregular narrowing of supraclinoid portion of both internal carotid and middle cerebral arteries, and numerous lenticulostriate and thalamic perforating collaterals giving "puff of smoke" appearance (Fig. 2A and B). These features were consistent with the diagnosis of cerebral moyamoya disease. DSA of abdomen revealed nonvisualisation of right renal artery with only a small stump seen opposite origin of left renal artery (Fig. 3A). DSA of pelvis showed short segment occlusion of right external iliac artery with distal reformation via collaterals from branches of internal iliac artery (Fig. 3B). MRI of abdomen confirmed the absence of right kidney (data not shown).

Hypertension was controlled with sodium nitroprusside infusion for 24 h, followed by oral clonidine, amlodipine and prazosin. Her seizures were controlled with anti-epileptic drugs. The child was started on oral aspirin (50 mg once daily). At discharge, the child's muscle power had improved to 4/5 and 3/5 on the right and left sides, respectively. Neurosurgical management has been planned in follow up.

3. Discussion

'Moyamoya disease' is a cerebrovascular disorder of unknown etiology characterized by progressive occlusion of supraclinoid internal carotid arteries and variable occlusion of its branches. When the similar clinical and radiological features are seen in association with conditions including Down's syndrome, neurofibromatosis, autoimmune disease, cerebrovascular atherosclerosis, cranial irradiation or cerebral neoplasms it is called "moyamoya syndrome". The presentation and associations of moyamoya disease are quite heterogenous. The commonest presentations in children include ischemic infarction, headache, and seizures [9]. Cerebral angiography is the gold standard for diagnosis of moyamoya disease. The angiography characteristically shows stenosis of terminal portions of internal carotid arteries with or without stenosis of middle and anterior cerebral arteries. The collateral vessels that form secondary to occlusions have characteristic 'puff of smoke' appearance. The diagnosis of moyamoya disease in our patient was based on the MRI, MRA and DSA findings. In addition, our patient also had cerebral atrophy, which has been described as a secondary phenomenon in moyamoya disease, occurring as a result of chronic ischemia [10].

Our case highlights two hitherto undescribed associations with childhood moyamoya disease: renal agenesis and external iliac artery occlusion. Renal involvement has been noted in moyamoya disease in the form of renal artery stenosis, renal artery aneurysm and nephrotic-range proteinuria; however renal agenesis has not been described before [2,11].

Involvement of peripheral arteries is a rare phenomenon in moyamoya disease. In a large series of 73 patients, none had stenosis of extra-renal abdominal arteries [3]. External iliac artery involvement has been reported in a single case report in a 30-year old woman [12], whose angiography had revealed diffuse luminal narrowing and smooth stenoses in right femoral bifurcation, left external iliac, and left superficial femoral arteries. Our patient also showed external iliac artery

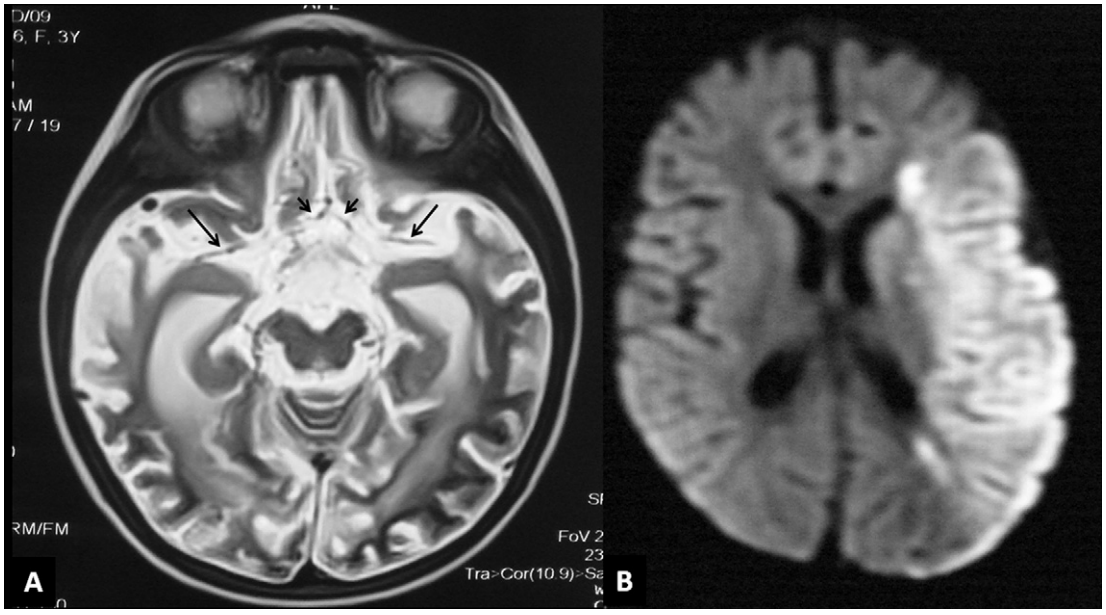

Fig. 1. Brain MRI examination. (A) T2 weighted axial imaging shows bilateral diffuse cerebral atrophy. The vascular flow voids of bilateral middle cerebral arteries (long arrows) and anterior cerebral arteries are attenuated. (B) Diffusion weighted axial image at a higher level shows acute infarct in left middle cerebral artery territory.