CASE REPORT

CHILDHOOD MOYAMOYA DISEASE WITH Ivy SIGN ON MRI

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ABSTRACT

A 2-year old girl referred to our department with complaint of transient ischemic attack and right sided hemiparesis, was investigated with contrast enhanced MRI brain and time of flight MRA and diagnosed as moyamoya disease with ivy sign. The ivy sign is a magnetic resonance imaging (MRI) finding observed both in post contrast T1-weighted images (T1WI) and fluid attenuated inversion recovery (FLAIR) images due to the development of leptomeningeal collateral and pial network. Moyamoya disease is a progressive multifocal occlusive arteriopathy of unknown origin and is an infrequent cause of cerebrovascular accident in children. The patient was treated conservatively and referred to a higher centre for specific neurosurgical revascularization surgery which has favorable outcome but because of his family disagreement cerebral revascularization surgery was not performed.

Key words: Moyamoya disease, ivy sign, leptomeningeal collaterals

Introduction

Moyamoya disease is defined as an idiopathic intracranial vascular disorder characterized by progressive stenosis/occlusion of internal carotid arteries termini/proximal anterior cerebral or middle cerebral arteries with subsequent development of multiple collaterals at the base of brain including lenticulostriate and thalamoperforators vessels as well as leptomeningeal and transdural anastomosis.[1,2] Moya Moya is a Japanese term, which means “hazy” derives its peculiar name from the angiographic appearance of cerebral vessels that resembles a “puff of smoke”.[3,4]

The ivy sign is a unique, characteristic magnetic resonance imaging (MRI) finding frequently observed in patients with moyamoya due to formation of leptomeningeal collaterals developed between external carotid and internal carotid arteries.[1] It can be observed both in post contrast T1-weighted images due to the enhancement of leptomeningeal collateral as well as increased numbers of pial vessels and in fluid attenuated inversion recovery (FLAIR) images due to slow arterial flow in the leptomeningeal collateral vascular channels.[1,5] In this case (approved by ERC of our Institute), we presented “ivy sign” of moyamoya disease on MRI in a 2 years old female patient. Additionally, we discuss its radiological features and review the literature. This case highlights the importance that early diagnosis and management leads to favorable outcome in children with Moyamoya disease, which is the purpose of reporting this case.

Case Report

A 2-year-old female child born of non-consanguineous marriage was admitted at our institute with complaints of weakness over left upper and lower limb for 10 days. There was also history of sudden fall 3 times probably indicating transient ischemic attacks. There was no history of fever, convulsion, head injury, bowel

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or bladder incontinency, loss of consciousness, eye deviation, tongue biting or ear discharge. There was no history of delayed mile stones, metabolic disorders or neurologic disease. There were no neurocutaneous markers or asymmetry of face. She was born full-term with vaginal delivery. No history of medical illness or neurologic disorder was reported in her parents and other family members. On neurological examination, gait was hemiplegic, tone was decreased over right side, power was 3/5 over left upper and lower limb, deep tendon reflexes were exaggerated, and planter was extensor over left side. Cranial nerve examination was within normal limit. Chest, heart and abdomen examination was within normal limits. Skin and mucous membrane did not reveal any unusual pigmentation, rash or pallor. Hematological examination including bleeding time, clotting time, prothrombin time, protein C & S were normal. Sickling test was negative. CSF analysis was unremarkable. Plain CT scan of brain was within normal limit. MRI brain showed T2 and FLAIR hyperintense signals in left basal ganglia with diffusion restriction and gyriform hyperintense signals along left fronto-parieto-temporal region in FLAIR images that showing contrast enhancement in post-contrast images (Ivy sign). Partial agenesis of corpus collasum was seen as an incidental finding (Fig. 1). MRA revealed non-visualization of supraclinoid segment of left internal carotid artery (ICA) and M1 segment of left middle cerebral Artery (MCA). Reduced caliber of right CCA, M1 as well as M2 segments of right MCA and A1 segment of right anterior cerebral artery (ACA) were also seen as compared to left side. Collaterals are seen around base of brain which gives "puff of smoke" appearance (Fig. 2), more on left side. Both posterior cerebral arteries (PCA) and posterior communicating arteries (PcoA) were prominent. Based on clinical and imaging findings the diagnosis of moyamoya disease was made. The patient was treated conservatively and was referred to higher centre for cerebral revascularization surgery as our centre is not equipped with such kind of pediatric neurosurgery. But because of his family disagreement cerebral revascularization surgery was not performed.

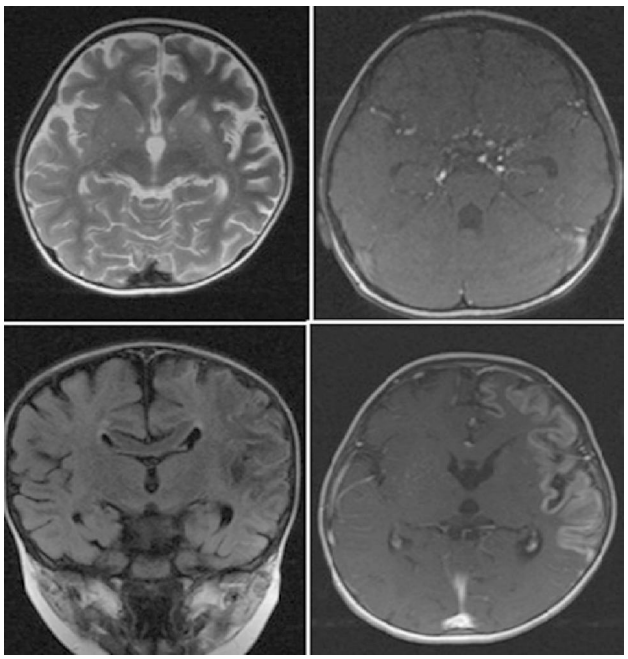

Figure 1: MRI brain showed infarct in left basal ganglia, collateral vessels around base of brain, Ivy sign along left fronto-parieto-temporal region in FLAIR and post-contrast images and partial agenesis of corpus collasum.

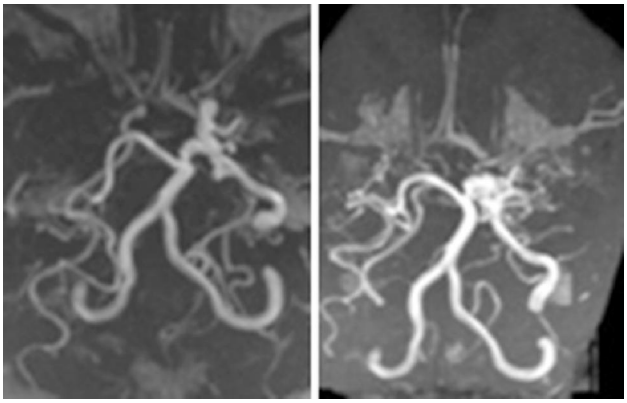

Figure 2: TOF- MRA revealed non-visualization of supraclinoid segment of left ICA as well as left M1 segment, reduced caliber of right CCA, right M1 as well as M2 segments and right A1 segment, collaterals giving "puff of smoke" appearance more on left side and prominent bilateral PCA as well as PcoA.

Discussion

Moyamoya disease is a non-atherosclerotic progressive occlusive arteriopathy of the distal internal carotid artery and the proximal middle cerebral as well as anterior cerebral arteries accompanied with development of multiple collaterals.[1,3] In Japanese, Moya Moya means "hazy" or "puff of smoke" due to cerebral vasculature appearance on angiography.[3,4] The condition is bilateral but asymmetric,[6] as seen in our patient. Unilateral involvement can occur in children in about 18% but usually within 2 years it progress to bilateral involvement.[1] Moyamoya disease

has been reported with bimodal age distribution showing high peaks at age 5 and low peaks at age 40,[6,7] and females are affected nearly twice as often as males,[2,8] our patient was also 2-year female. Clinical manifestations of moyamoya disease differ in children and adults. In children it most commonly presents with transient ischemic attacks with infarcts predominantly in the ICA territory, may accompanied with monoparesis, hemiparesis, aphasia and dysarthria, our patient was also presenting with these manifestations while in adults the subarachnoid and/or intra parenchymal bleeding is the prevailing presentation.[2,4]

MRA is the best imaging modality to confirm diagnosis of moyamoya disease with sensitivity of 73% and specificity of 100% but when MRA is combined with MRI brain the sensitivity can be increases to 92%.[8] MRA shows the occlusion and narrowing of distal internal carotid arteries or proximal part of cerebral vessels with collateral arteries while MRI shows an infarct in ICA territory and ivy sign,[2,3] similar findings were seen in our patient's MRA and MRI brain. The 'Ivy sign' was first described in 1995 in post contrast MR sequences but later on, it was also reported in FLAIR images, due to formation of leptomeningeal collaterals developed between external carotid and internal carotid arteries seen in approximately 70% of moyamoya patients.[1,5,7] It was named as ivy sign due to resemblance with ivy creeping on stones.[5,7] The "ivy sign" refers to diffuse leptomeningeal and cortical enhancement due to formation of collaterals in post contrast MR images and leptomeningeal and cortical hyperintensity due to slow arterial flow on FLAIR imaging.[1,5] The gold standard imaging method for the diagnosis and surgical planning of moyamoya disease is conventional cerebral angiography especially for visualization of smaller collaterals. However, since cerebral angiography is invasive and not easy to perform, particularly in paediatric age groups as well as due to risk of radiation exposure, non-invasive MRI and MRA are the primarily preferred imaging methods.[7]

Acute management is supportive but long-term favorable outcome is achievable with revascularization surgery.4 So our patient was managed conservatively and referred to a higher centre for specific neurosurgical revascularization surgery but because of his family disagreement cerebral revascularization surgery was not performed.

Conflict of Interest: None

Conclusion

The diagnosis of moyamoya disease is now easily achievable with modern neuroimaging techniques (MRI and MRA), especially in children. It is important to be familiar with the clinical and MRI/MRA findings in moyamoya disease to make an early diagnosis. Since early diagnosis and intervention will alter the prognosis and life expectancy especially in pediatric patients. Conventional cerebral angiography remains the gold standard for diagnosing and surgical decision making for patients with suspected moyamoya disease.

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