RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

Case Report: A rare case of Moyamoya Disease

KEYWORDS

* Dr. Saurabh Chaudhuri	Dr. Priscilla Joshi
Associate Professor, Department of Radiodiagnosis and Imaging, Bharati Hospital and Research Centre, Pune, * Corresponding Author	Professor and Head, Department of Radiodiagnosis and Imaging, Bharati Hospital and Research Centre, Pune
Dr. Mohit Goel	**Dr. Wasim Siddiqui**
Resident, Department of Radiodiagnosis and Imaging, Bharati Hospital and Research Centre, Pune	Resident, Department of Radiodiagnosis and Imaging, Bharati Hospital and Research Centre, Pune

ABSTRACT *This case documents an unusual case of Moyamoya disease in a 5 year old girl who presented with chief complaint of seizures since two years. MR angiography reveals stenosis and irregularity of the both supraclinoid ICAs and non-visualization of bilateral MCAs and ACAs with extensive basal collateralization giving the characteristic "puff of smoke" appearance consistent with Moya Moya.*

INTRODUCTION:
"Moyamoya" is a Japanese word meaning puffy, obscure, or hazy like a puff of smoke in the air. Moyamoya disease is an idiopathic chronic progressive cerebrovascular disease characterized by bilateral stenosis or occlusion of the arteries around the circle of Willis with prominent arterial collateral circulation. Collateral circulation from the extracranial arteries to the intracranial arteries includes ethmoidal moyamoya from the ophthalmic artery and transdural anastomoses derived from the middle meningeal and superficial temporal arteries. The incidence of moyamoya disease among females is 1.8 times greater than among males and the highest incidence is found in Asia, especially Japan (0.35–2.0 cases per 100000; prevalence 3.16) [1].

CASE REPORT:
A 5 year old female child presented with history of seizures since two years which has increased in frequency since few days. MRI was performed on PHILIPS 1.5 T ACHIEVA scanner. Multiplanar, multiecho MR of the brain was performed before and after administration of IV gadolinium. T1, T2 and FLAIR images were obtained in different planes. MR angiogram and MIP images were also obtained.

FLAIR hyperintensities are noted in bilateral fronto-parietal convexity sulci (left > right) showing post-contrast enhancement [Figure 1], could represent Ivy sign. MR angiography reveals stenosis and irregularity of the both supraclinoid ICAs and non-visualization of bilateral MCAs and ACAs with extensive basal collateralization giving the characteristic "puff of smoke" appearance consistent with Moya Moya [Figure 2]. Few abnormal focal hyperintense signals were also noted in the fronto-parietal subcortical white matter on FLAIR and T2WI. No diffusion restriction and GRE blooming was noted.

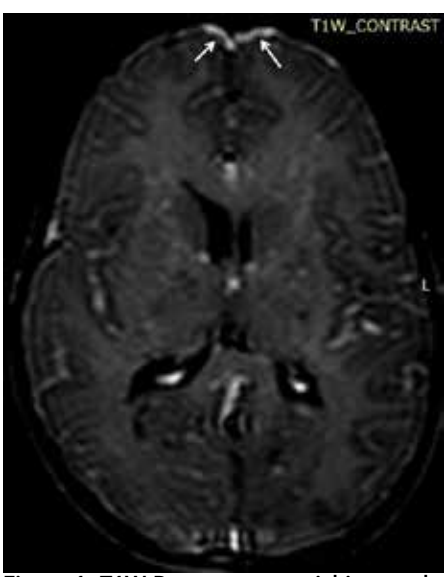

Figure 1: T1W Post contrast axial image shows leptomeiningeal enhancement in bilateral fronto-parietal convexity sulci (left > right) - Ivy sign.

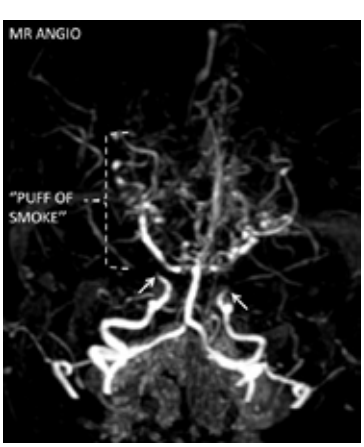

Figure 2: MR angiography reveals stenosis and irregular-

Volume : 4 | Issue : 4 | Apr 2014 | ISSN - 2249-555X

ity of the both supraclinoid ICAs (arrows) and non-visualization of bilateral MCAs and ACAs with extensive basal collateralization giving the characteristic "puff of smoke" appearance.

DISCUSSION:

Moyamoya disease is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels seen on imaging, particularly cerebral angiography [2]. Moyamoya has two peak ages of presentation. Two thirds of cases occur in children and one third in adults with peak presentation in the fifth decade.

Clinically, the disease may be of ischemic, hemorrhagic and epileptic type. Cognitive dysfunction and behavioral disturbance are atypical symptoms of moyamoya disease [3]. When moyamoya vessels are associated with an underlying condition such as connective tissue disease, the condition is referred to as Moyamoya Syndrome.

Imaging plays an important role in establishing the diagnosis. MR imaging is a good technique for screening for Moyamoya Disease. MR angiography is used to confirm the diagnosis and to see the anatomy of the vessels involved. It can reveal stenosis or occlusion of the supraclinoid internal carotid artery and basal moyamoya vessels by signal voids as well as ischemia, infarction, atrophy, and ventricular dilatation. The ivy sign is referred to the diffuse leptomeningeal enhancement that resembles creeping ivy on stones in patients with Moyamoya Disease. The ivy sign can be found on FLAIR images, as well as on contrast-enhanced MR images [4].

However conventional angiography is the standard diagnostic technique in patients with Moyamoya Disease and needs to be performed for accurate staging. Suzuki and Kodama classified the evolution of Moyamoya Disease using angiographic findings to define six phases of the disease:

Stage I: Narrowing of carotid fork; stage.
Stage II: Initiation of basal moyamoya.
Stage III: Intensification of moyamoya.
Stage IV: Minimization of moyamoya.
Stage V: Reduction of moyamoya.
Stage VI: Disappearance of moyamoya.

Early diagnosis and treatment are important during childhood as the disease can cause progressive disability, including deterioration in cognitive ability, [5] because viable cerebral tissue is necessary for reperfusion. For children and adults with moyamoya and acute stroke, acute treatment is mainly symptomatic and directed towards reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures [6].

Revascularization surgery using direct and indirect bypass provides effective surgical management for pediatric Moyamoya Disease. However, surgical treatment of the adult hemorrhagic type remains controversial. Rehabilitation with physical therapy, occupational therapy, and speech therapy should be considered, depending on the neurologic impairment.

REFERENCE

1. Wetjen NM, Garell PC, Stence NV,et al. Moyamoya disease in themidwestern United States. Neurosurg Focus 1998; 5:e1. | 2. Janda PH, Bellew JG, Veerappan V. Moyamoya disease: case report and literature review. J Am Osteopath Assoc. 2009 Oct; 109(10):547-53. | 3. Eugeniusz Tarasów et al. Moyamoya disease: Diagnostic imaging. Pol J Radiol. 2011 Jan-Mar; 76(1): 73–79. | 4. Maeda M, Tsuchida C. "Ivy sign" on fluid-attenuated inversion recovery images in childhood moyamoya disease. AJNR Am J Neuroradiol 1999; 20:1836-1838. | 5. Khan N, Schuknecht B, Boltshauser E, et al. Moyamoya disease andMoyamoya syndrome: experience in Europe; choice of revascularization procedures. Acta Neurochir (Wien) 2003;145: 1061–71. discussion 1071. | 6. Fukui M. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ('moyamoya' disease). Research Committee on Spontaneous Occlusion of the Circle of Willis (Moyamoya Disease) of the Ministry of Health and Welfare, Japan. Clin Neurol Neurosurg 1997; 99 Suppl 2:S238. |