

Moya Moya disease: Report of 2 cases and review of literature.

Showkat MS¹, Nuruzzaman S², Datta A³

Abstract :

Moya moya disease is a chronic cerebral vasculopathy characterized by progressive occlusion of circle of Willis arteries that leads to development of collateral vessel which has a characteristic puff of smoke appearance on imaging. Here we described 2 cases, both were young and presented with history of stroke and repeated headache. In Magnetic Resonance Angiogram (MRA), we found marked narrowing in internal carotid, anterior, middle and posterior cerebral artery with multiple collaterals producing puff of smoke appearance which is the diagnostic imaging feature of Moya Moya disease. This is an important cause of repeated stroke in young patients. So physicians should be aware of this clinical entity.

➤ CBMJ 2013 July: Vol. 02 No. 02 P: 78-80

Key words: Moya moya, Cerebral vasculopathy, repeated headache, smoke appearance on imaging,

Introduction:

Moya Moya disease is a chronic cerebral vasculopathy first described in 1957 by Taceuchi and Shimizu from Japan, who named the diseases in 1969.¹ In Japanese Moya Moya means 'hazy'. The disease derived its unusual name from angiographic appearance of cerebral vessels that resembles a 'puff of smoke'.² This chronic cerebrovascular disease is characterized by progressive occlusion of circle of Willis arteries leading to the development of characteristic collateral vessels seen on imaging, particularly in cerebral angiography.³ Clinically, it represents with cerebral ischemic or haemorrhagic events. Ischemic events occur mainly in childhood while haemorrhagic events occur in adulthood. Thus Moya Moya disease has a bimodal incidence with severe morbidity and mortality. Incidence of this disease among female is 1.8 times high than among males and the highest incidence is found in Asia especially in Japan.⁴ Here two cases of Moya Moya disease has been described and literature about this disease has been reviewed.

Case Reports:

Case 1:

A 5 years old young girl was admitted to the Neurology department of Bangabandhu Sheikh Mujib Medical University (BSMMU) with the complaints of right sided hemiparesis and unconsciousness for 10 days. She had previous

history of repeated headache which was throbbing in nature.

The patient came to Radiology and Imaging Department for Magnetic Resonance Imaging (MRI) and MRA of brain. MRI of brain showed- Sub acute infarct at left capsulo ganglionic region. Old infarct of left frontal periventricular region and anterior part of centrum semiovale along with white matter ischemic changes. MRA of brain showed marked narrowing of supraclinoid portion of both internal carotid arteries and proximal part of both anterior and middle cerebral arteries. Extensive collateral vessels were seen around anterior part of circle of willis producing puff of smoke appearance. Moderate luminal narrowing was seen at P1 and P2 segment of both posterior cerebral arteries.

1. *Dr Mst Syeeda Showkat
Assistant Professor, Dept of Radiology & Imaging,
Bangabandhu Sheikh Mujib Medical University
(BSMMU), Dhaka, Bangladesh.
2. Dr Sheikh Nuruzzaman
Radiologist, Green Life Medical College and Hospital
Dhaka, Bangladesh.
3. Dr Anindita Datta
MD(Resident), Dept of Radiology and Imaging,
Bangabandhu Sheikh Mujib Medical University
(BSMMU). Shahbag, Dhaka, Bangladesh.

* **Address of correspondence:**
E-mail : drjany2008@yahoo.co.uk
Mobile : 01550002276



Figure 1: Findings of MRA of brain in case 1

Case 2:

A 19 years old female patient was admitted to the Neurology Department of BSMMU with the complaints of fever, headache, irrelevant talk and right sided hemiparesis. The patient was referred to Radiology and Imaging Department for MRI and MRA of brain. MRI of brain showed acute infarcts at left occipito temporal and parietal region with old infarcts at both parietal and right temporal regions. Numerous prominent perivascular spaces of both capsulo ganglionic regions due to extensive collateral vessels was also evident. MRA showed marked luminal narrowing at terminal part of both internal carotid arteries, arterial circle of willis and whole length of anterior and middle cerebral arteries. Moderate luminal narrowing was also noted at whole length of Posterior cerebral artery and extensive collateral vessels were noted at both capsulo ganglionic region producing puff of smoke appearance.

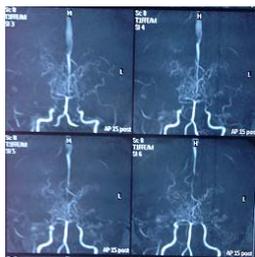


Figure 2: Findings of MRA of brain in case 2

Both this patient were young and presented with history of stroke and repeated headache. In MRA, we found marked narrowing in internal carotid, anterior, middle and posterior

cerebral artery with multiple collateral producing puff of smoke appearance which is the diagnostic imaging feature of Moya Moya disease.

Discussion:

Moya Moya disease is a rare cerebral vasculopathy of unknown etiology which is characterized by progressive narrowing of major intracranial vessels and development of collateral vessels. Previously it was thought that moya moya disease is prevalent only in japanese individuals but now it has been found in all races with varying age distribution and clinical manifestation.^{4,5} However, majority of the cases are reported in Asia and other non caucasian regions⁶.

The process of narrowing of cerebral vessels seems to be a reaction of brain blood vessels to a wide variety of external stimuli, injuries or genetic defects². Primary Moya Moya disease is inherited polygenic or autosomal dominant disease with low penetrance. Secondary Moya Moya disease is associated with other diseases such as sickle cell anaemia, Neurofibromatosis-1, Down's syndrome, a congenital heart defects, antiphospholipid syndrome, renal artery stenosis, thyroiditis⁷. The process of blockage once begins; it tends to continue despite any known medical management unless treated with surgery⁸.

Presenting features are usually different in Adult and children. Adults usually present with sign and symptoms of TIAs, cerebral haemorrhage or infarct. Haemorrhagic presentation is more common in Asian adults. Best diagnostic tools for Moya Moya disease are MRI and MRA. MRI reveals area of infarction as well as visualization of collateral vessels as multiple dots like flow voids at base of the brain and basal ganglia in T1 weighted image. In T2 weighted scan increased signal of small vessel cortical and white matter infarcts are seen. Net like cisternal filling defect of collateral vessels are also seen. MRA is used to confirm the diagnosis and to see the anatomy of vessels involved. It typically reveals the narrowing and occlusion of distal internal carotid artery (ICA)

and proximal common carotid artery (CCA) together with extensive collateral flow through

the perforating vessels, demonstrating the classic puff of smoke appearance⁹.

Acute management of Moya Moya disease is mainly symptomatic and aim is to reduce elevated Intra cranial pressure, to blood flow and to control seizures. Surgical revascularization procedures are definite mode of treatment.

Direct bypass to superficial temporal artery–middle cerebral artery (STA–MCA) is more common in adult and indirect bypass by encephalo-duro-arterio-synangioses (EDAS) is more effective in children. Hemodynamic improvement after surgical procedures appears to be similar in all age groups¹⁰.

References:

1. Takeuchi K, Shimizu K. Hypogenesis of bilateral Internal carotid artery. *Shinkei* 1957; 9: 37-43.
2. Goyal JP, Rao SS, Trivedi S. Moya Moya disease in a child: A case report. *Case reports in Neurological Medicine*. 2011. ID 329738.
3. Janda PH, Bellew JG, Veerappan V. Moya Moya disease. *J Am Osteopath Assoc*. 2009 ;109(10);547-53.
4. Wetjen NM, Garell PC, Stence NV. Moya Moya disease in the Midwestern United States. *Clinical Neurology and Neurosurgery*. 1998; 99(2):s31 – s35.
5. Khan N, Schuknecht B, Boltshauser E. Moya moya disease and Moya moya syndrome : experience in Europe; choice of revascularisation procedures. *Acta Neurochirurgica*.2003.145;12: 1061-1071.
6. Natori Y, Ikezaki K, Matsushima T, Fukui M. Angiographic Moyamoya; its definition, classification and therapy. *Clinical Neurology and Neurosurgery*.1997;99(2): s168- s172.
7. Lutterman J, Scott M, Nass R, Geva T. Moyamoya syndrome associated with congenital heart disease. *Pediatrics*. 1998;101(1): 57-60.
8. Smith ER, Scott RM. Moyamoya syndrome associated with congenital heart disease. *Skull base*. 2005;15(1): 15-26.
9. Atlas SW, MR *Angiography: Techniques and clinical Applications MRI of the Brain and Spine*. Lippincott Williams and Wilkins, Philadelphia, USA, 3rd edition. 2002.
10. Han DH, Kwon OK, Bark BJ. A cooperative study : clinical characteristics of 334 Korean patients with Moyamoya disease treated at neurosurgical institutes (1976- 1994) *Acta Neurochirurgica*.2000;142(11): 1263- 1274.