MOYA-MOYA DISEASE IN A CHILD - A CASE REPORT

Shivshankar D¹*, Pushpalatha S¹, Reddy CN¹

1. Bangalore Medical College and research Institute. India

Correspondence
Dr. Shivshankar Diggikar. Bangalore Medical College and research Institute. India
Email: shiv.diggikar@gmail.com


ABSTRACT

We report a case of five-year-old female child who was admitted in our hospital with complaints of right sided hemiparesis and aphasia. On MR angiography, the child was diagnosed to have Moya- Moya disease. Moya-Moya disease is a rare cause of cerebral stroke in children. In our case as the age of onset was very early and it marks poor prognosis, suggesting that early surgical intervention is needed. The patient was treated conservatively and referred to a higher center for specific neurosurgery. Neurosurgical revascularization procedures leads to favorable outcome.

Key words: Moya-Moya disease, report

INTRODUCTION

We report a case of five-year-old female child who was admitted in our hospital with complaints of right sided hemiparesis and aphasia. On MR angiography, the child was diagnosed to have Moya- Moya disease. Moya-Moya disease is a rare cause of cerebral stroke in children. In our case as the age of onset was very early and it marks poor prognosis, suggesting that early surgical intervention is needed.

CASE REPORT

A five years old female child presented with weakness of right upper and lower limb, deviation of face towards left side, inability to speak since eight days, child also had incontinence of urine and stools. There was history of tuberculosis two years back (father was a K/C/O tuberculosis), took treatment adequately for six months. Family history was not significant.

On examination: There was right sided hemiparesis with facial weakness. The sensory system was normal with no meningeal or cerebellar signs. Other systemic examination was normal.

Hematological investigations were normal (complete blood count/LFT/RFT/ESR), except child had moderate anemia with Hb of 8gm%. CSF analysis was normal (not suggestive of tuberculosis), 2D ECHO was normal. MRI contrast brain revealed Multifocal acute
lacunar infarct in left frontal, left gangliocapsular, left temporal, left temporo-parietal and left temporo-frontal region. Chronic lacunar infarct in left centrum semiovale. Old infarct with gliosis and atrophy in right temporal lobe, right temporo-occipital, right frontal and fronto-parietal white matter. Bilateral white matter ischemic changes noted. MRI- Angio of carotid and intracranial arteries revealed smooth narrowing and occlusion of pre cranial part of right internal carotid artery. Long segment smooth narrowing and tapering with complete occlusion of distal pre cranial part of left internal carotid artery. Multiple Moya-Moya collaterals are seen around perimesencephalic and the circle of Willis region giving rise to smoky appearance. On the basis of MRI features suggestive of Moya-Moya disease, probable Moya-Moya disease was diagnosed. There was no systemic features. We treated the child with Aspirin (3mg/kg/d). Child showed mild improvement in power 3/5. As our center was not equipped with neurosurgical intervention, we referred the child to higher center for encephaloduroarteriosynangiosis.

Figure 1. MRI finding. (LEFT) MRI ANGIO showing narrowing of left internal carotid artery with distal collaterals giving "PUFF OF SMOKE" appereance .(RIGHT) MRI CONTRAST showing multiple lacunar infarct.

DISCUSSION

Moya - Moya disease is a rare disease characterized by multiple occlusions of the cerebral circulation with an unusual net like system of collaterals. In Japanese, Moya-Moya means “hazy”. The disease derives its peculiar name from the angiographic appearance of cerebral vessels in the disease that resembles a “puff of smoke”. In children, the most common presentation is that of recurrent episodes of cerebral ischemia manifesting clinically as focal deficits, paresthesia, and seizures [1]. Previously thought
to be prevalent only in Japan, cases have now been reported from across the globe. However, majority of the cases are reported in Asia and other non-Caucasian regions [2]. 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 defects. Conditions such as sickle cell anemia, neurofibromatosis-1, Down's syndrome, congenital heart defects, antiphospholipid syndrome, renal artery stenosis, and thyroiditis have been found to be associated with Moya Moya disease in the literature [3]. MRI not only reveals areas of infarctions, but also allows direct visualization of these collateral vessels as multiple small flow voids at the base of the brain and basal ganglia. MR angiography is used to confirm the diagnosis and to see the anatomy of the vessels involved. It typically reveals the narrowing and occlusion of proximal cerebral vessels and extensive collateral flow through the perforating vessels, demonstrating the classic puff of smoke appearance [4]. Treatment is complex. Antiplatelet is often employed and is safe. Carbonic anhydrase inhibitor may shift acid–base balance to favor vasodilatation. In severe Moya-Moya such medications may cause a ‘steal’ or ‘reverse robin hood phenomenon [5]. Revascularization procedures are currently performed to increase the perfusion to the hypoxic brain tissue. The literature supports these procedures, and long-term favorable outcome has been reported in terms of improvement in symptoms and positive angiographic follow up in all age groups [6].

This case highlights the importance that though early age of onset has poor prognosis, but early diagnosis and intervention helps in improving the overall outcome.

CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare that they have no competing interests.

REFERENCES