East African Scholars Journal of Medical Sciences
Abbreviated Key Title: East African Scholars J Med Sci
ISSN: 2617-4421 (Print) & ISSN: 2617-7188 (Online)
Published By East African Scholars Publisher, Kenya

OPEN ACCESS

Volume-3 | Issue-6| June-2020 | DOI: 10.36349/EASMS.2020.v03i06.004

Case Report

Moya Moya disease in children

Dr. Hitender Gautam[1], Dr. Surender Singh[2], Dr. Priyanka[3*], Dr. Amit Sachdeva[4]

[1]Junior Resident, Department of Paediatrics, Indira Gandhi Medical College, Shimla, Himachal Pradesh
[2]Associate Professor, Department of Paediatrics, Indira Gandhi Medical College, Shimla, Himachal Pradesh
[3]Junior Resident, Department of Community Medicine, Indira Gandhi Medical College, Shimla, Himachal Pradesh
[4]Senior Resident, Department of Community Medicine, Indira Gandhi Medical College, Shimla, Himachal Pradesh

Article History
Received: 21.05.2020
Accepted: 09.06.2020
Published: 15.06.2020

Journal homepage:
https://www.easpublisher.com/easjms

Quick Response Code

Abstract: Moya Moya Disease (MMD) is a rare cerebrovascular disorder of unknown aetiology. It consists of stenosis or occlusion of main cerebral arteries which causes an abnormal vascular network at the base of the brain, which appears like puff of smoke on MR angiography. Here we report 3 cases of Moya Moya disease presenting to our institute Indira Gandhi Medical College, Shimla over a period of one year (2017-18) including 1 case of MMS with beta thalassemia .Possibility of Moya Moya disease should be kept in patients with stroke and motor deficit. Gold standard to diagnose the disease is conventional angiography. There is no definitive medical treatment to reverse or stabilize the course of MMD. Early diagnosis and intervention like revascularize surgeries will alter the prognosis.

Keywords: Moya Moya Disease , cerebrovascular disorder, stenosis

INTRODUCTION

Moya Moya (MMD) disease is a rare cerebrovascular disorder of unknown aetiology. It consists of stenosis or occlusion of main cerebral arteries which causes an abnormal vascular network at the base of the brain, which appears like puff of smoke on MR angiography. (Suzuki, J., & Takaku, A. 1969). The disease was first described by Takeuchi and Shimizu *et al.* in Japan (1957). This disease presents in children as stroke and seizures. It was first described in Japan where its incidence is as high as 6.03/ lakh population. Globally its Incidence is 0.5 /lakh population (Fujimura M & ToMinaga T, 2015). Although bilateral involvement is typical of MMA, angiographically proven unilateral steno-occlusion of terminal ICA has also been involved in some cases (Sanefuji M *et al.,* 2006). Moya Moya disease associated with other recognized diseases, such as meningitis in childhood, neurofibromatosis type-II, Down syndrome, cranial irradiation and different types of anaemia, particularly hemoglobinopathies are also known as Moya Moya syndrome (MMS) . Although sickle cell disease is a well-described risk factor for MMS (Marden FA *et al.*, 2008) there are only few cases of MMS associated with thalassemia (Vernet O, *et al.*,1996). Here we report 3 cases of Moya Moya disease presenting to our institute Indira Gandhi Medical College, Shimla over a period of one year (2017-18) including 1 case of MMS with beta thalassemia.

CASE REPORTS

Case 1: 12 year male child was diagnosed case of beta thalassemia major transfusion dependent for last 10 years admitted with weakness in left upper limb, deviation of face to right side for last 2 days. Child also had history of sudden onset weakness in both lower limbs 10 days prior to admission had complete recovery after 30 minute. There is no history of seizure any injury. Family history is also not contributory

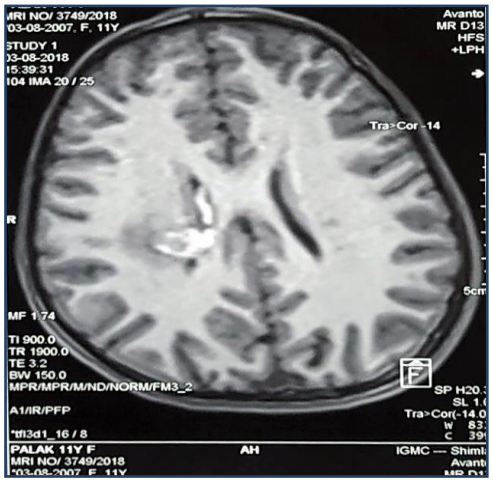
On examination the child was conscious oriented. Power in left upper limb was 3/5 and normal in rest of the limbs. Cranial nerve examination shows 7th nerve palsy on left side UMN type. All haematological investigation was normal except Hb which was 6.8gm/dl. MRI brain shows acute infarct in Pre motor cortex right frontal lobe and right peri-ventricular region, MR angiography show attenuation in right MVA and ICA finding suggestive of Moya Moya

***Corresponding Author:** Dr. Priyanka

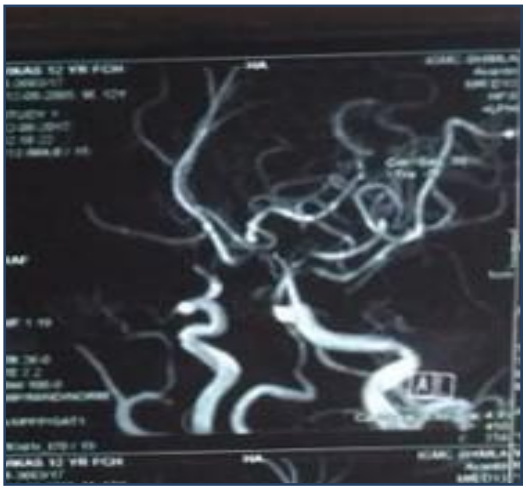
disease. Child was managed conservatively on aspirin. Follow up was done for 1 year during this period child had no complaint.

Case 2: 3year old female child admitted with h/o multiple episodes of generalized tonic clonic seizures (GTCS) from last 10 days. There was no h/o fever, rash, drug intake, ear discharge and injury .Her father also revealed that she had sudden weakness in left side of body one year back which was improved over a period of 6-8 months .on examination her vitals are normal and systemic examination was not significant, no focal deficit at the time of examination, All haematological investigations were normal. MRI findings are suggestive of Moya Moya disease. This child was also managed conservatively with antiepileptic**.**

Case 3: 11year female admitted with chief complains of weakness in left hand and headache for three days, there was no h/o fever, seizures, head injury and drug intake. She was diagnosed case of spastic diplegia with hypomylenosis 4 years back on MRI in 2014.On examination she was conscious oriented with power of 3/5 with increase tone in left upper limb and extensor planters. Rest all examination was normal. All haematological investigations were normal. MR angiography findings were multiple collateral along bilateral basal ganglion and along anterior circulation with attenuated supraclinoid part of bilateral ICA suggestive of Moya Moya disease. This patient was started on asprin and discharged.



MR image-1


MR image-2

DISCUSSION

Moya Moya disease is a chronic, progressive occlusion of the main cerebral artery that leads to the development of characteristic collateral vessels seen on cerebral angiography. In children, the most common presentation is that of recurrent episodes of cerebral ischemia manifesting clinically as focal deficits, paraesthesia, and seizures (Scott RM, *et al.*, 2009). 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" (Lutterman J, *et al.*, 1998).

Etiopathogenesis of Moya Moya disease is poorly understood. 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 anaemia, 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 (Taher AT *et al.*, 2010). Hypercoagulability, followed by thromboembolic events, is a widely recognized complication of thalassemia which lead to recurrent emboli in attenuated vessels (Lutterman J *et al.*, 1998). Family history of thrombotic event, previous splenectomy, profound anaemia, and a serum ferritin level≥1000 mg/l are few risk factors for thrombosis in bête thalassemia. On the other hand, positive history of transfusion and a haemoglobin level≥9 g/dl were found to be protective against thrombosis. The process of blockage, once it begins, tends to continue despite any known medical management unless treated with surgery (Atlas, S.W, 2002). MR angiography 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. Acute management is mainly symptomatic and directed towards reducing elevated intracranial pressure, improving cerebral blood flow, and controlling seizures, Surgical revascularization is thought to improve cerebral perfusion, and to reduce the risk of subsequent stroke. Prognosis of Moya Moya disease is found to be better in younger age . TIA and epileptiform clinical pictures have a better long-term outcome (Inoue TK, *et al.*, 2000).

CONCLUSION

Moya Moya disease is a rare cerebrovascular disease. Possibility of Moya Moya disease should be kept in patients with stroke and motor deficit. Gold standard to diagnose the disease is conventional angiography. There is no definitive medical treatment to reverse or stabilize the course of MMD. Early diagnosis and intervention like revascularize surgeries will alter the prognosis.

REFERENCES

1. Suzuki, J., & Takaku, A. (1969). Cerebrovascular moyamoya disease: disease showing abnormal net-like vessels in base of brain. *Archives of neurology*, *20*(3), 288-299. Takeuchi K, Shimizu K. Hypogenesis of bilateral internal carotid arteries [inJapanese]. No To Shinkei. 1957;9:37-43.
2. Kim, S. K., Wang, K. C., Kim, D. G., Paek, S. H., Chung, H. T., Hee, M., ... & Cho, B. K. (2000). Clinical feature and outcome of pediatric cerebrovascular disease: a neurosurgical series. *Child's Nervous System*, *16*(7), 421-428.
3. Fujimura, M., & ToMinaga, T. (2015). Diagnosis of moyamoya disease: international standard and regional differences. *Neurologia medico-chirurgica*, ra-2014.
4. Dobson, SR., Holden, K.R., Nietert, P.J., Cure, J.K, Laver, J.H., Disco, D. (2002). Moyamoya syndrome in childhood sickle cell disease: a predictive factor for recurrent cerebrovascular events. *Blood*, 99, 3144–3150.
5. Sanefuji, M., Ohga, S., Kira, R., & Yoshiura, T. (2006). Moyamoya syndrome in a splenectomized patient with β-thalassemia intermedia. *Journal of child neurology*, *21*(1), 75-77.
6. Marden, F. A., Putman, C. M., Grant, J. M., & Greenberg, J. (2008). Moyamoya disease associated with hemoglobin Fairfax and beta-thalassemia. *Pediatric neurology*, *38*(2), 130-132..
7. Vernet, O., Montes, J. L., O'Gorman, A. M., Baruchel, S., & Farmer, J. P. (1996). Encephaloduroarterio-synangiosis in a child with sickle cell anemia and moyamoya disease. *Pediatric neurology*, *14*(3), 226-230.
8. Scott, R. M., & Smith, E. R. (2009). Moyamoya disease and moyamoya syndrome. *New England Journal of Medicine*, *360*(12), 1226-1237.
9. Lutterman, J., Scott, M., Nass, R., & Geva, T. (1998). Moyamoya syndrome associated with congenital heart disease. *Pediatrics*, *101*(1), 57-60.
10. Cappellini, M. D., Poggiali, E., Taher, A. T., Musallam, K. M., Weatherall, Rund, ... & Cappellini. (2012). Hypercoagulability in β-thalassemia: a status quo. *Expert review of hematology*, *5*(5), 505-512. Taher AT, Otrock ZK, Uthman I, Cappellini MD .Thalassemia and hypercoagulability. Blood Rev.2008; 22:283–292.
11. Taher, A. T., Musallam, K. M., Karimi, M., El-Beshlawy, A., Belhoul, K., Daar, S., ... & Cappellini, M. D. (2010). Overview on practices in thalassemia intermedia management aiming for lowering complication rates across a region of endemicity: the OPTIMAL CARE study. *Blood, The Journal of the American Society of Hematology*, *115*(10), 1886-1892.
12. Lutterman, J., Scott, M., Nass, R., & Geva, T. (1998). Moyamoya syndrome associated with congenital heart disease. *Pediatrics*, *101*(1), 57-60.
13. Atlas, S. W. (2002). MR angiography: Techniques and Clinical Applications MRI of the Brain and Spine.
14. Inoue, T. K., Ikezaki, K., Sasazuki, T., Matsushima, T., & Fukui, M. (2000). Linkage analysis of moyamoya disease on chromosome 6. *Journal of child neurology*, *15*(3), 179-182.