INTRODUCTION

Moya Moya (MMD) disease is a rare cerebrovascular disorder of unknown etiology. 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 revascularise surgeries will alter the prognosis.

**Keywords**: Moya Moya Disease, cerebrovascular disorder, stenosis.

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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 aspirin and discharged.

**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 bêta 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