Role of Physiotherapy Intervention in Rare Case of Moya Moya Syndrome - A Single Case Study

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Abstract

Introduction: Moya moya syndrome is a moya moya angiopathy related secondarily to any neurological symptom or because of a specific acquired or inherited origin presenting with sensory and motor impairments. Although the disease is quite common in Japan, many cases have been reported around the globe, including North America, Europe, and India. Epidemiologic studies in Japan have estimated the annual incidence of moya moya disease to be 0.35 to 0.94 per 100000 people. This case studies the role of physiotherapy as it is not explored much in the paediatric population.

Material and Methodology: A child who was diagnosed with moya moya syndrome presented to us at the age of 3.3 years and was evaluated on GMFM-88 scale, Visuo-Motor Integration (VMI) component of PDMS-2 scale along with Manual Ability Classification System (MACS). Physiotherapy intervention was done based on impairments to improve motor function.

Results: GMFM-88 was administered for 4 times in 18 months. A total score of 38 was observed at first administration and a total of 24 score was observed at 18th month of administration. The VMI score of PDMS-2 scale did not change pre and post intervention.

Conclusion: This single case study shows about the nature of the syndrome and individualized integrated protocol which is needed for the evaluation and tretment in the cases of moya moya syndrome population which needs to be segregated from cerebral palsy.

***Keywords**: Moya moya syndrome, GMFM-88, VMI, MACS, Physiotherapy.*

Introduction

Moya moya disease (MMD) was first described in 1957 by Takeuchi and Shimizu in Japan.[1] "Moya moya disease" for the first time was coined by Suzuki and Takaku in 1969.[2] Although the disease is most common in Japan, several subsequent cases have been reported in North America, Europe, and India as well.[3,4,5] Epidemiologic studies in Japan have estimated the annual incidence of moya moya disease to be 0.35 to 0.94 per 100000 people.[6,7] In the present literature, the incidence and prevalence rate of this disease and syndrome in India is not yet known.[8]

Moya moya disease is a chronic progressive non-atherosclerotic, non-inflammatory and non-amyloid cerebrovascular disorder which is defined as progressive stenosis of the intracranial vessels.[9,10] MMD is a cerebrovascular disease which is rare and progressive in nature, characterized by bilateral occlusion and

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abnormal formation of collateral vessels substituting internal carotid artery and branches forming the circle of Willis.[11,12] Moya moya syndromes (MMS) correspond to moyamoya angiopathy which is amalgamated with further neurological or extra-neurological presentations, with or without an inherited or an acquired condition.[8] The stenosis begins from the terminal bifurcation of internal carotid arteries and gradually progresses to the anterior, middle, and posterior cerebral arteries.[10,13]

Children present most frequently with transient ischemic attacks or ischemic strokes with acute infantile hemiplegia. Headaches, involuntary choreiform movements and motor disturbances are the most common mode of presentation occurring in 80.5% of the population. Convulsions occur in about 9%.[14]

In the literature there is an inadequate amount of knowledge about physical rehabilitation in MMD. Generally, the studies have mostly focused on applications and complications of revascularization surgery techniques.[15] For this purpose, this single-case study reports the changes in motor function and functional ability of a child with Moya moya syndrome, over a clinical observation. Moya moya syndrome is rare and role of physiotherapy is not broadly explained as it is explained in other condition like cerebral palsy as literature is limited on this topic. Hence, this post operative case is taken to study the role of physical therapy intervention and study the course of syndrome.

Case description

A 3.3 years old male presented to us with a complaint of no head holding since two years and was diagnosed with moya moya syndrome by the paediatric neurologist. When the child presented to us his height was 92 cms, weight was 13.5 kgs and BMI was 15.9kg/m^2. The child was apparently normal until 8 and a half months of age and was said to have achieve all the milestones. The first episode of convulsion lasted for 3 seconds with upward gaze and clenching of teeth. There was a history of two more episodes of epilepsy at 9 and a half month and 11 months of age respectively.

Both the plantar reflexes were up going. The child presented to us with exaggerated reflexes present in the upper and lower limbs. Also, the tone in the upper and lower limb muscles was increased which was assessed on Modified Ashworth Scale. The tone in the biceps was 1 on the right side whereas on the left it was 1+, in the hip extensors it was 2 bilaterally, in the knee flexors and plantar flexors it was 3 bilaterally.

The result of brain MR imaging showed subacute infarct in right Middle cerebral artery (MCA) territory, lacunar infarct in right frontal region and gliosis due to old vascular insult in right basal ganglia. Also, there were acute non- hemorrhagic infarcts in left fronto-parietal lobes in parafalcino region, genu and anterior limb of left internal capsule in left Anterior cerebral artery(ACA) territory with tiny acute infarct in right frontal white matter and right lentiform nucleus infarct. According to the child's medical record he underwent a surgery for bilateral pterional craniotomy and bilateral encephalo-duro-arterio-myo-synangiosis (EDAMS) with bilateral frontal burrhole. The MRI and MRA of our child showed classical internal carotid artery stenosis and infarct of the brain tissue.

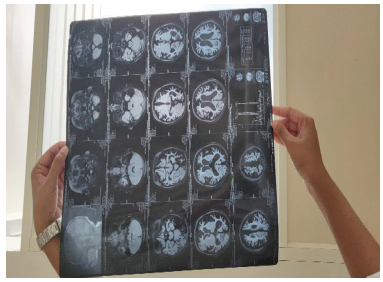

Figure - 1

1) MRI showing atrophy of parenchyma

The child is currently on Clobazam, Baclofen, Amantadine hydrochloride, Cholecalciferol capsule, Ferrous ascorbate, Folic acid and Mecobalamin suspension.

Post surgery when the child was stable he was brought to us for further physical rehabilitation. Informed consent was taken from the parent before initiating the rehabilitation and the child was evaluated on GMFM-88, VMI component of PDMS-2 and also on MACS level. The therapy was given for a total of 45-60 minutes for 3-4 days a week. The intervention consisted of visual stimulation, neck holding facilitation, creeping facilitation, rolling facilitation and hand weight bearing which was given for 7-10 minutes each with oral motor facilitation given for 3-5 minutes. Treadmill walking was done along with all these exercises for 10 minutes.

Home exercises were taught to the parent. The present developmental stage of the child showed intermittent neck holding in the gross motor domain, fine motor activity was not achieved, there was a severe impairment in the cognition level of the child. In the speech and language domain, child had achieved cooing whereas in the visual domain, he could not fix nor track the light and was able to feed on semi solid foods.

A)

B)

Figures-Intervention protocol pictures

Table 1: GMFM SCORES

Items	COMPONENTS	I ADMINIS-TRATION	II ADMINIS-TARTION	III ADMINIS-TRATION	IV ADMINISTRATION
A	LYING AND ROLLING	27	36	36	15
B	SITTING	11	12	14	9
C	CRAWLNG	0	0	0	0
D	STANDING	0	0	0	0
E	WALKING	0	0	0	0
	TOTAL SESSIONS		Between I and II administration-52	Between II and III administration-12	Between III and IV administration-35

TABLE 1- The participant of the current study had been following with us for 18 months and was assessed on GMFM-88 and VMI component of PDMS-2 for this current study. The number of sessions completed between I & II administration were 52 and the score improved from 38 to 48 (+10) which showed 3.79% gain. Whereas between II & III administration the sessions were 12 and the score improved from 48 to 50 (+2) which showed a gain of 1.51%. From III & IV administration there was an increase in the number of sessions upto 35 but the score deteriorated from 50 to 24 (-26) wherein we had a loss of -9.84%. Therefore, the net score was found to be 14 for 99 sessions when calculated between the first (38) and the final administration (24). The VMI score of PDMS-2 component scale when administered; pre intervention score was zero and post intervention the score did not change.

Discussion

The prognosis of MMD may present with gradual, fulminate, intermittent attacks or rapid loss of neurological signs.[11,12]

In the previous literature, GMFM-88 was used to assess the gross motor function as it is a gold standard method. The child who was of 4 years of age at the time of his diagnosis was rehabilitated for 2 years and showed about a 100% improvement for his functional recovery in his gross motor function. The GMFM-88 scores were assessed every month.[14] In another study wherein the child was of 10 years of age. The manual function test (MFT) was adopted for evaluation of upper limb movement and hand manipulative function in each segmental joint of the affected limb. In addition, the child's ambulatory ability was tested using the functional ambulatory category (FAC).[15] In the previous studies both the children had not undergone surgery whereas in this study the child had undergone bilateral EDAMS surgery.

In our study the total number of sessions were 99 and the child was assessed on GMFM-88 on regular basis wherein we found a change in score initially but later the scores did not change and this could be contributed towards the illness of the child, the nature of the progression of the disease along with multiple system involvement of cognitive and visual system.

Also, in our study we have considered VMI component of PDMS-2 scale wherein the score remained zero pre and post intervention. Besides this we have included MACS level of the child and the child falls in level V of this classification system. Although the same classification system was not considered in the previous literature.[14,15]

As per the literature, this is one of the very few studies that investigated the role of physiotherapy program in MMS. Previous studies have described the role of MMS in adult population[16,17,18] and thus the role of MMS ought to be explored more in the paediatric population. Along with the diagnostic methods and surgical applications, it should be accentuated that further studies should be done on physical therapy assessment and applications with appropriate sample size and randomized clinical trials.[14, 15]

As physiotherapy rehabilitation may improve the functional status and reduce the disabilities in MMS, this population is under more risk from other childhood neurological disability such as Cerebral palsy and other neurodevelopmental disorders. In addition, MMS have a similar representation as cerebral palsy and the cases are treated for the same as spastic quadriplegic cerebral palsy. Hence, this component should be taken into consideration for physiotherapy management as it is one of the main approaches. And this study describes the role of physiotherapy in moya moya syndrome.[14]

In contrast to the previous studies, this single case, studies the regression in development that may be due to multi-system deficits like cognitive, visual, motor and other barriers in rehabilitation like regularity in therapy.[14,15]

Conclusion

In conclusion, this study gives us a guidance that as our child had visual and cognitive impairment, there was no much improvement seen. As moya moya syndrome is progressive in nature, the therapist should be aware that it may not show the outcome as seen in non progressive disease. Hence, maintenance therapy plays a major role.

Conflict of Interest: None

Source of Funding: Self

Further scope: 1) Case series.

2) Longer follow-up.

3) To study the prevalence and incidence of Moya Moya Syndrome in Indian scenario.

Ethical Clearance- Patient informed consent was taken from the parents for publication and all the other norms were followed.

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