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EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Functional recovery in a child with Moyamoya disease

Moyamoya hastalığında fonksiyonel iyileşme

Mehmet Duray[1], Arzu Genç[2]

[1]Pamukkale University, School of Physical Therapy and Rehabilitation, Denizli, Turkey
[2]Dokuz Eylül University, School of Physical Therapy and Rehabilitation, İzmir, Turkey

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Dear Editor

Moyamoya disease (MMD), 'puff of smoke' in Japanese is a rare but progressive cerebrovascular anomaly characterized by bilateral stenosis and occlusion of the arteries that comprise Willis circle. Collateral vessels can be observed with cerebral angiogram. Moyamoya disease is usually seen with ischemic cerebrovascular strokes in children while it is derived from hemorrhage in adults. After strokes, neurological sequels usually appear such as transient ischemic attacks, motor deficits, sensory disorder, involuntary movement, recurrent headaches, seizures and cognitive disorders in pediatric patients. Due to recurrent strokes, severe functional disorders and death can be seen, even so early identification and treatment of the lesions is vital. In the literature there is inadequate amount of knowledge about physical rehabilitation in MMD generally the studies have mostly focused on applications and complications of revascularization surgery techniques[1,2,3].

A 4-years-old child was diagnosed as having Moyamoya Disease and was rehabilitated for 2 years. She had been born at the 37th week of gestational age. Physical examination at the born revealed: weight, 2670 g; height, 47cm; head circumference, 34 cm. Her mother reported having a history of chrioamnionitis. When she was 10-month-old, she had febrile convulsion in the form of whole body tonic clonic contractions for 3-5 minutes. After 1,5 months; one night, while crawling she had fallen her right side. She had not cried, couldn't move her eyes and mouth for 5 minutes. Neurological examinations of deep tendon reflexes, eye movements and MRI results were normal at the hospital. When she was fourteen month MRI, taken from routin control, showed bilateral occlusion of both of anterior and middle cerebral arteries. In subsequent MRI analysis; atrophy of frontal, parietal and partially temporal region, dilatation of cortical sulcus, lateral and third ventricles, subdural hemorrhage and Wallerian degeneration at mesencephelon were observed. Then she was diagnosed as having MMD.

The parents of the child gave their written informed constent to participation in this case report. The study protocol was approved by the ethics committee of Dokuz Eylul University Hospital (Izmir, Turkey).

Age, height, body weight, orthotic usage was recorded. Modified Ashworth Scale (MAS) was used to determine the presence and severity of spasticity of bilateral upper and lower extremity muscles. Scale was composed of 5 items[4]. (0 = normal muscle tone, 4 = limb rigid in flexion or extension). Gross motor function was evaluated with Gross Motor Function Measurement-88 (GMFM-88) during 8 months. GMFM is gold standard for clinical applications and rehabilitation researches. GMFM-88 is composed of 5 motor domains and 88 items. Motor domains of GMFM-88 are lying and rolling; sitting; crawling and kneeling; standing; and walking, running, and jumping[5]. Physical Therapy Program had included combination of spasticity control training in

Yazışma Adresi/Address for Correspondence: Dr. Mehmet Duray, Pamukkale University, School of Physical Therapy and Rehabilitation, Denizli, Turkey E-mail: mehmetduray@gmail.com
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different positions, balance and weight bearing training, strengthening exercises, vestibular stimulation and sensory training. Program was applied one time in a week with games. Home program was given to the family and educated about disease and how to apply exercise at home. So patient is obligated to exercise at home every day. The physiotherapy program was progressed according to the changes in the patient's clinical status.

The patient had been followed for 8 months and physical examination was done every month. During treatment the patients did not attack. 4 years old this child's height was 99 cm, weight was 37.8 kg. Her parents were not relatives. She has been using Ankle Foot Orthosis, orthopedic shoes and walker. Total GMFM-88 scores for each month from January (baseline) to August were 28.49, 29.82, 34%, 37.4, 41.79, 44.79, 46.96 and 49.97, as % respectively (Table 1). Gross motor function of our patient had shown improving about 100%. The spasticity was evaluated by using MAS according to muscle groups the severity of spasticity is demonstrated in Table 2. Especially reducing the spasticity in the plantar flexor muscles takes attention.

MMD is a rare, progressive cerebrovascular disease, characterized by bilateral occlusion and developmental caused abnormal collateral vessels in internal carotid artery and branches forming Willis circle[1,2]. The prognosis of MMD may show variety, may be slow, fulminate, intermittent attacks or rapid neurological loss. Repetitive cerebral ischemia and intracranial hemorrhage causes hemodynamic changes[1,2].

Table 1. GMFM scores of patient on every month

	January	February	March	April	May	June	July	August
GMFM-88 Scores	28.49	29.82	34	37.4	41.79	44.79	46.96	49.97

Table 2. Muscle tone assessment

Muscle Groups	**Modified Ashworth Scale Scores**			
	Pre treatment		**Post treatment**	
Lower Extremity	Right	Left	Right	Left
Hip				
Flexors/Extensors	0/0	0/0	0 / 0	0/0
Adductors/Abductors	0/0	1/0	0 / 0	1/0
Internal/External Rotators	0/0	1/0	0 / 0	1/0
Knee				
Flexors/Extensors	0 / 0	0/1	0 / 0	0/1
Ankle				
Plantar flexors/Dorsi flexors	1 / 0	3/0	1/0	2/0
Invertors/Evertors	1 / 0	2/0	1/0	1/0
Upper Extremity	R	L	R	L
Shoulder				
Flexors/Extensors	0 / 0	1 / 0	0 / 0	1 / 0
Adductors/Abductors	0 / 0	1 / 0	0 / 0	1 / 0
Internal/External Rotators	0 / 0	1 / 0	0 / 0	1 / 0
Forearm				
Flexors/Extensors	0 / 0	2 / 0	0 / 0	2 / 0
Pronators/Supinators	0 / 0	0 / 1	0 / 0	0 / 1
Wrist				
Flexors/Extensors	0 / 0	2 / 0	0 / 0	2 / 0

Symptoms usually occurs depending on hemorrhage in adult and ischemia in children[1,6]. Despite MMD can be seen in all the races in the World, it is more common in Japanese and Korea and can be observed in women two times more than men[2,3]. The familial story is stated as 7-12% in Japanese[3]. Symptoms caused by ischemia were seen in our case. But the familial history had not been represented.

There isn't any current treatment method that can stop the progression of the MMD[1]. Initial treatment aims to maintain cerebrovascular function with symptomatic treatment methods[4,5]. However, it is

believed that revascularization surgery, categorized as direct, indirect and combined bypass, is useful in pediatric patients Revascularization surgery is executed (implement) for nearly 18% of patients with unilateral MMD[2,6,7]. Our 4 years old case with unilateral MMD hadn't been implemented revascularization surgery.

In younger patients, progressive vascular occlusion causes repetitive ischemic attacks[4]. In children, if uniteral occlusion has seen, other side effects within 1-2 years[3]. At first, right side of our case has influenced, then the left side was influenced too. However, our case had the appearance of left side hemiplegic.

Functional, visuospatial perception, organization and construction impairments, motor and sensory problems, involuntary movements, recurrent headaches, seizures, and neurologic symptoms such as cognitive disorders may be seen in MMD[6]. In some cases affected posterior circulatory may lead to ischemic ataxia, vision problems and vertigo may be observed[3]. Speech loss, facial sagging, arm weakness and ataxic movements were also reported in the literature[4]. In our case, spasticity, upper and lower motor movement limitation because of spasticity, intermittent headaches, seizures history and losing of speech ability were observed.

GMFM; is used as gold standard for determining gross motor function changes in pediatric patients who have movement and function disability such as Cerebral Palsy[4]. Nam et al. reported motor development on 10-years-old case with MMD after 3 years application, that consisted of medical therapy and physiotherapy program with strengthening exercises, functional activity training and neurodevelopmental treatment[8]. Similarly, we focused on motor function and improvement of 4 years old case with MMD and we prefer GMFM for gross motor function. Within eight months, we have seen functional level had risen from 28.49% to 49.97%.

Physiotherapy aims to develop motor function in children with developmental disorder like childhood stroke. as soon as possible these children should be included the rehabilitation programs for motor and neurological recovery. After eight months follow-up physiotherapy program, our case gained independency in supine and prone position approximately and could stand on her knees. Especially in sitting position, it was observed that symmetric body perception increased during weight bearing and upper extremity movements. while our case couldn't stand before treatment, she succeeded standing assisted after treatment. coleman et al. reported early inpatient rehabilitation was effective even if the patients with mmd are under two years old[14].

Papavasilou pointed that static weight bearing is used for enhancing antigravity muscle strength, reducing spasticity and improving hand functions, strengthening exercises are used for enhancing weak antagonist muscles against the spastic agonist muscles in children with CP10. In our study, we found that spasticity of plantar flexors and invertors decreased with multi factorial applications included spasticity control training, balance and weight transfer training, muscle strengthening, vestibular stimulation and sensory training.

According to our literature knowledge, this is the one of the pioneer studies that investigated the effectiveness of physiotherapy program in MMD. In addition to the diagnostic methods and surgical applications, it should be emphasized that further studies should be done on physiotherapy assessment and applications with comprehensive simple size.

In conclusion, we determined that physiotherapy applications may improve the functional status and decrease the disabilities in MMD. So this population that is under more risk from other childhood neurological disability such as CP, physiotherapy should be considered the one of main approaches.

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