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Role of Neuromuscular Electrical Stimulation and Physiotherapy in Managing Hirayama Disease: A Case Report

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Abstract

Background: Hirayama disease (HD) causes progressive weakness and atrophy in forearm and hand muscles on one side without sensory disturbances. There are no studies exploring the role of neuromuscular electrical stimulation (NMES) of intrinsic muscles of hand and physiotherapy in managing HD. In this report, we present a case of HD affecting the right arm and referred for physiotherapy.

Case Report: A 17-year-old male with right-hand weakness was diagnosed with HD. Initially, he experienced weakness in his middle and little fingers and muscle twitching in his forearm. The diagnosis was confirmed through a flexion-extension MRI of the C-spine, which revealed abnormalities in the laminodural space between C3 and C6. Physiotherapy was recommended to treat the condition. The patient showed muscle atrophy in the intrinsic muscles of the hand and underwent a 6-week physiotherapy program involving NMES and hand exercises. This stimulation was given at extensor digitorum, abductor pollicis longus and brevis, abductor digiti minimi, dorsal and palmar interossei. 50 contractions were given at each point. The patient also underwent gripping and prehension exercises with theraputty and theraweb. Hand function was assessed using Disability of the Arm, Shoulder, and Hand (DASH) questionnaire and Nine-Hole Peg Test (NHPT). The patient's DASH score improved by 10 points and was clinically significant.

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Conclusion: NMES and Physiotherapy can be useful for hand function in HD.

Keywords: Hirayama disease, Physiotherapy, Therapeutic electrical stimulation

INTRODUCTION

Hirayama and colleagues first reported Hirayama's disease (HD) in 1959. It is also known as juvenile muscular atrophy of the distal upper extremity (JMADUE) or monomelic amyotrophy. While it is less common among people in North America and Europe, the majority of reported cases of this illness come from Japan and other Asian nations (1,2,3).

The clinical diagnostic criteria involve observing discernible weakening and atrophy of the muscles of the distal forearm and hand, primarily affecting the thenar eminence, hypothenar area, and interosseous musculature. The brachioradialis muscle is notably spared, which results in the identifiable appearance known as "oblique amyotrophy" on one upper extremity side. The condition usually manifests itself between the ages of 10 and 20. Stretch reflexes are normal and there are no sensory problems; nonetheless, in order to provide an accurate diagnosis, additional disorders must be ruled out (3,4). The illness is self-limiting, first exhibiting a progressive phase that lasts for one to five years, then stabilizing for a while (5).

Although the exact etiology of the illness is unknown, one theory attributes spinal cord compression to an unbalanced growth spurt. This could cause the dura mater to shift during neck flexion, which could compress the spinal cord and cause anterior horn cell necrosis as well as disturbed microcirculation. The unique characteristics of HD can be shown by flexed cervical spine MRI (6). Studies have shown that Neuromuscular Electrical Stimulation (NMES) therapy can significantly reduce severe hand impairment in individuals with chronic stroke which are sustainable (7). Studies examining the function of physiotherapy and NMES in the treatment of HD do not exist. We describe in this report a case of HD that affected the right arm and was sent for physiotherapy.

CASE REPORT

The informed consent form was signed and the institutional Ethics in Research Committee authorized the current report. A male 17-year-old right hand dominant patient arrived at the physiotherapy department in November 2022 with progressive right-hand weakness that affected his grip and writing. His neurologist had referred him. March 2020 marked the onset of his middle finger weakness, which was followed by forearm fasciculation and the incapacity to elevate the ring and little fingers. He also observed that bending of the neck resulted in the flexion of fingers in the right hand. The patient disclosed a history of using a phone in a supine position on a sofa with his head resting on the armrest and a frequent neck-flexed posture. He used to play volleyball during COVID-19 restrictions and observed arm fasciculation.

When he saw a neurologist in November 2020, the doctor recommended that he get an MRI of his brain, cervical spine (C-spine), and nerve conduction velocity (NCV) testing. The ulnar nerve axonal neuropathy was indicated by longer latency and lower compound muscle action

potential (CMAP) amplitude in the motor NCV obtained for the right abductor digiti minimi below the elbow (Table 1).

Table 1. Details of the motor nerve conduction study

Nerve/Sites	Muscle	Latency (ms)	Amplitude (mV)	Amplitude (%)	Duration (ms)	Segments	Distance (cm)	Latency Difference (ms)	Velocity (m/s)
R Median Nerve -APB									
Wrist	APB	3.33	0.0	-	5.68	Wrist-APB	NR	NR	NR
Elbow	APB	11.25	-	-	3.85	Elbow-Wrist	21	7.92	32
L Median Nerve - APB									
Wrist	APB	4.79	8.0	93.4	7.03	Wrist-APB	-	-	-
Elbow	APB	9.69	8.6	100	7.34	Elbow-Wrist	21	4.90	51
R Ulnar Nerve - ADM									
Wrist	ADM	3.44	6.0	100	7.34	Wrist-ADM	7	-	-
Below Elbow	ADM	9.95	5.3	88	8.07	Below Elbow- Wrist	23	6.51	43
Above elbow	ADM	10.89	5.1	83.9	8.18	Above Elbow- Below Elbow	10	0.94	53
L Ulnar Nerve – ADM									
Wrist	ADM	4.43	4.2	100	7.03	Wrist – ADM	7	-	-
Below Elbow	ADM	9.84	4.1	97.8	6.98	Below Elbow- Wrist	23	5.42	52
R Radial Nerve - EIP									
Forearm	EIP	2.81	2.2	100	7.86	Forearm –EIP	-	-	-
Elbow	EIP	5.57	2.0	91.8	8.28	Elbow-Forearm	-	2.76	-
Spiral Groove	EIP	7.66	2.1	93.4	8.80	Spiral Groove-Elbow	-	2.08	-
R Axillary Nerve - Deltoid									
Erb's Point	Deltoid	3.70	0.7	100	13.39	-	NR	NR	NR
L Axillary Nerve - Deltoid									
Erb's Point	Deltoid	3.28	1.3	100	6.77	-	-	-	-

R Right; L Left; APB Abductor Pollicis Brevis; ADM Abductor Digiti Minimi; EIP Extensor Indicis Proprius; NR Not recordable; ms millisecond; mV millivolt; m/s meter per second; cm centimeter

The MRI revealed no noteworthy abnormalities. Sensory NCV was normal. Patient was given a tentative diagnosis of HD and instructed to take multivitamins, baclofen 5 mg, and wear a soft cervical collar. He went to another neurologist in September 2022 for a second opinion, and this one suggested a flexion-extension MRI of the C-spine. The posterior thecal sac was effaced as a result of aberrant laminodural space enlargement in the flexion position between the C3 and C6 levels. The cord contour flattened as a result of dural indentation on the posterior chord surface. When the neck was in neutral extension, these findings disappeared. Both flexion and extension MRI showed that the cervical central canal was normal, and there was no evidence of disc herniation or foraminal stenosis. There were no anomalies in the anterior atlanto-axial articulation. There were no signs of tonsillar herniation. When the NCV test was conducted again, no unusual results were found. Following confirmation of the HD diagnosis, physiotherapy was recommended thereafter. Upon physical examination, there was wasting of the right medial forearm and atrophy of the thenar, hypothenar, and interossei muscles. The muscle brachioradialis was unaffected (Figure 1). On being instructed to extend all the fingers the patient was unable to extend the middle and ring finger on the affected side (Figure 2). The right arm's muscle strength in the dorsal interossei, abductor pollicis longus and brevis, abductor digiti minimi, and extensor digitorum was found to be 2/5 by the Medical Research Council (MRC). The right triceps received a 3/5 rating. The left arm received a 4/5 MRC rating. The

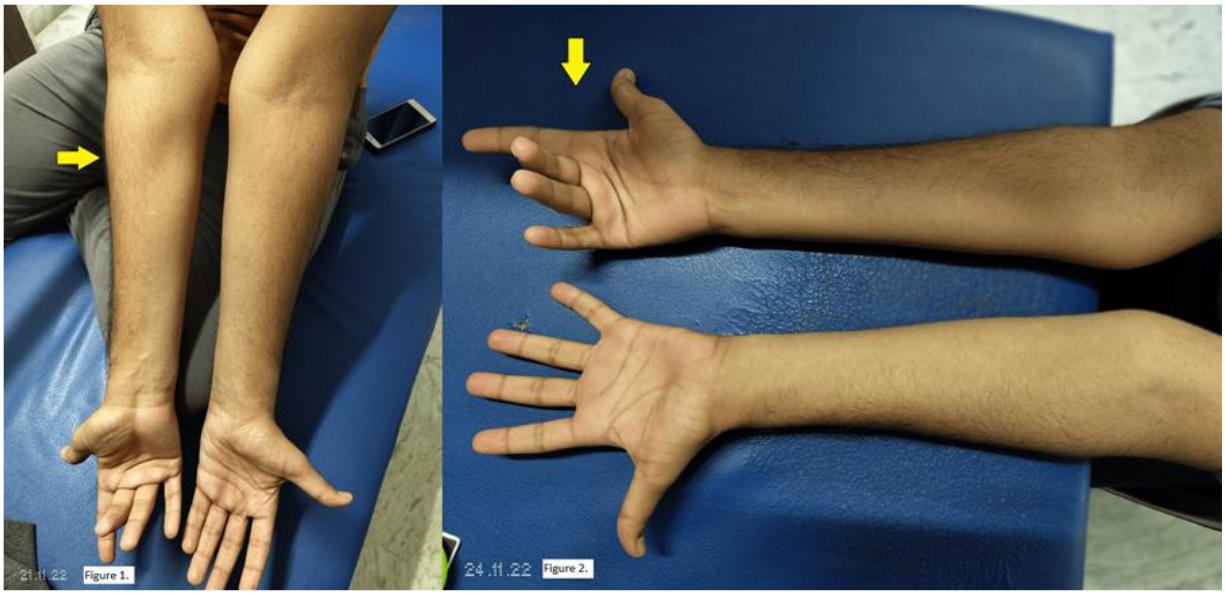

Figure 1. With both forearms in supination, noticeable muscle atrophy is observed on the medial side of the right forearm. The brachioradialis muscle, located in the same region is spared.

Figure 2. With both forearms resting in a supinated position on a table, when prompted to voluntarily extend all fingers, the patient demonstrates an inability to extend the middle and ring fingers on the right side.

patient did not report any eyelid drooping, double vision, pain, sensory loss, or difficulties swallowing. Furthermore, there was no family history of such medical conditions, and the patient's coordination and gait were normal. The muscles of the lower limbs were unaffected.

Treatment intervention

The 12-week, six-day-a-week program included motor point stimulation of the hand's intrinsic muscles, grasping exercises for the hands, and NMES of the finger extensors employing surged faradic current. The cathode of NMES was positioned on the belly of the extensor digitorum, while the anode was positioned at the forearm's common extensor origin. The NMES lasted for about thirty minutes in total. Faradic current with pulse duration of 100 millisecond at 100 Hz was used to stimulate motor points. The abductor digiti minimi, dorsal and palmar interossei, extensor digitorum, and abductor pollicis longus and brevis were the targets of this stimulation. At every motor point, fifty contractions were administered. Additionally, the patient practiced gripping and prehension with theraweb and theraputty. With the theraweb, the patient places fingers through the openings, performing controlled opening (extension) and closing (flexion) movements against resistance and presses the entire hand to mimic a gripping motion. Using theraputty, the patient squeezes it with the entire hand, holding for 5-10 seconds and repeating 10-15 times. The patient also pinches small sections with the thumb and fingertips, rolls the putty into a ball or cylinder, and shapes it to enhance fine motor skills. Additionally, the patient presses the thumb into the putty, forming a dimple, to improve thumb function and coordination. Using 1 kg dumbbells, supine elbow extension exercises were done for 15 repetitions over three sets.

We used the Disability of the Arm, Shoulder, and Hand (DASH) questionnaire, which includes scoring in 29 out of 30 areas, to assess arm function. Minimal Clinically Important Difference (MCID) approach was used to to check for the clinically significant improvement in the DASH scores. MCID of 10 point is considered significant (8). The DASH score at baseline was 42.5; following the 12-week intervention period, it improved to 32.5. The finger dexterity was measured using the Nine-Hole Peg Test (NHPT). A Minimal Detectable Change (MDC) approached was used for NHPT with MCD of 4 and 7 seconds being significant for dominant and non dominant hand respectively (9). For the right and left hands, the baseline scores on the NHPT were 29 and 24 seconds, respectively. The scores for the right and left hands increased to 22 and 20 seconds, respectively, after the 12-week PT intervention.

RESULTS

The patient's functional capacities significantly improved after the 12-week PT session. The patient's ability to undertake everyday tasks was evident as the DASH score decreased from 42.5 to 32.5 with an overall change in 10 points. The

NHPT scores showed better finger dexterity, going from 29 to 22 seconds for the right hand and from 24 to 20 seconds for the left.

Improvements were also seen in the patient's muscular strength. The right hand's muscle strength increased to a 3/5 in the dorsal interossei, abductor pollicis longus and brevis, abductor digiti minimi, and extensor digitorum, using the MRC grading system. The right triceps received a rating of 4/5. The left arm's MRC rating stayed at 4/5.

DISCUSSION

HD is a benign disease that results in "oblique amyotrophy," or focused muscle wasting, mainly in the C7, C8, and T1 myotomes while sparing the brachioradialis muscle. It mostly affects the muscles of the upper limbs innervated by C5-6 myotomes. The illness, which mostly affects young boys, causes progressive muscle atrophy and weakness that frequently goes away on its own. A constricted canal and stretched cord result from an imbalance in the growth of the dura and vertebrae. Because of the disruption of microcirculation in the anterior region of the spinal cord during neck flexion, the anterior shift of the dura compresses the spinal cord and may result in ischemic injury (10).

Reverse split hand syndrome is a unique pattern associated with HD that is commonly revealed by nerve conduction investigations. The abductor digiti minimi exhibits a drop in CMAP amplitude, or it is absent, in this pattern, whereas the abductor pollicis brevis retains its response. This unique characteristic differentiates HD from amyotrophic lateral sclerosis, which commonly manifests as the traditional split hand syndrome (11,12).

In a neutral neck position, a routine MRI usually reveals no abnormalities. On the other hand, it may show aberrant curvature or lower cervical cord atrophy. Conversely, during flexion MRI, notable observations could be the dural sac shifting forward, separating from the surrounding lamina, and a crescent-shaped mass appearing in the lower cervical canal's posterior epidural region. This mass is usually linked to posterior internal vertebral venous plexus congestion, which frequently goes away when the neck returns to its neutral posture (13).

A cervical collar worn pro-actively for three to four years effectively stops the advancement of HD, which usually has a self-limiting course lasting one to five years. About the role of physiotherapy in the management of disease, there is only anecdotal evidence available. There isn't any literature currently available about the benefits of PT and NMES in the treatment of HD. The DASH score increased by 10 points in our study, which is regarded as a minimally clinically significant variation. The NHPT results also revealed a notable improvement.

CONCLUSION

In summary, the main treatment for HD is a cervical collar, which stops the disease's progression and prevents neck flexion. The prognosis for HD patients is typically good.

Although physiotherapy has been shown to improve hand function and reduce handicap in activities of daily living, its contribution to meaningful therapeutic gains is still uneven. The patient in this case study experienced considerable functional improvements as a result of the application of NMES and hand therapy. The DASH and NHPT score improvements imply that these therapies may be useful in the management of HD. To assess the long-term advantages and the effect of these therapies on the restoration of motor function, muscle strength, and coordination in the afflicted hands and forearms, more research is necessary.

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Conflict of Interest Statement: The authors declare that they have no conflict of interest.

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