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Research Article

Hirayama's Disease- A Case Study

Dr Roopa Rao[1*], Dr Asawari Thakur[2], Dr Himanshu Sharma[3]

[1*]Assistant Professor, Dept of Physiotherapy, BKL Walawalkar College of Physiotherapy, Sawarde
[2]Intern, Dept of Physiotherapy, BKL Walawalkar College of Physiotherapy.
[3]Associate Professor, Dept of Physiotherapy, Mahalaxmi College of Physiotherapy and Rehabilitation Centre, Raigaon.

***Corresponding Author:** Dr Roopa Rao
*Assistant Professor, Dept of Physiotherapy, BKL Walawalkar College of Physiotherapy, Sawarde
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Introduction:

Hirayama disease, a condition with male preponderance also known as monomelic amyotrophy was first described by Hirayama in 1959. Highest prevalence of this condition is noted in Asian population with most cases reported from Japan. Cases have also been reported from other parts of Asia and few from European countries. It is a form of motor neuron disease that has an insidious onset and presents as unilateral or bilateral muscle atrophy and weakness without sensory loss.[1] This condition usually progresses for one to two years before plateauing. There are chronic ischaemic changes taking place in the anterior horn cells of the cervical region, especially in the C7 and C8 regions leading to myelopathy.[1] Imbalance between the vertebral column and dural canal during growth spurt is postulated to be the cause.[1,5] This imbalance causes increased laxity causing anterior displacement of posterior dura during flexion causing mechanical cord compression.[1,5] Even though the disease is non-progressive, management of symptomatic myelopathy is required. The use of a cervical collar and prevention of neck flexion is the first line of treatment.[1] If conservative methods fail, surgical intervention may be considered.[1] A study by YeTian reported that there was an association between posterior cervical extensors and cervical kyphosis. They concluded that weakness of posterior cervical extensors (deep and superficial) leads to poor cervical stability which was crucial in the management of HD.[8] The focus of this study is cervical muscle strengthening and core strengthening to improve spinal alignment.

Patient Information:

A 38-year-old male, a mechanic by profession, presented to the physiotherapy outpatient department with complaints of neck pain and weakness in his right hand. He complained of an inability to perform activities like turning a key, fixing screws at his work, and slipping objects from his hand after holding them for a little while. He was however able to do his ADLs with the assistance of a non-dominant hand. He also complained of the onset of neck pain for 3 years. The onset of weakness was insidious and the patient first noticed it when he was 23 years old. The condition seemed to be progressing due to which he consulted many physicians. He was diagnosed with Hirayama Disease (HD) in July 2017 based on clinical findings and electrodiagnostic reports. He was prescribed vitamin B12 supplements as supportive medicines. Apart from this his past medical history as well as family history was not significant. Informed consent was taken by us for further evaluation and reporting purposes. A detailed history was taken which revealed that the patient faced difficulties in his occupation due to the weakness of his dominant hand and had to switch the nature of his job as advised by his physician.

Clinical Findings:

On observation, there was evident atrophy of the thenar, hypothenar, and interosseus muscles of the right hand.

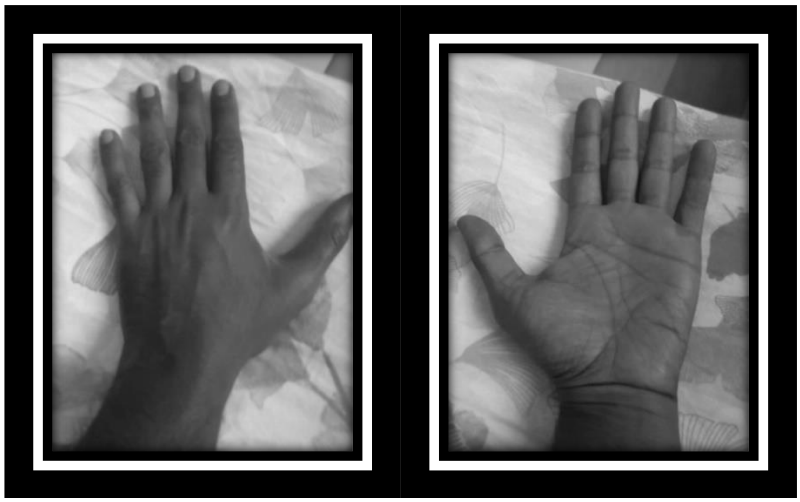

Figure 1 (Left Hand)- This shows thenar and hypothenar muscles atrophy. The atrophy is more on the ulnar side than the lateral side suggestive of reverse split hand syndrome.

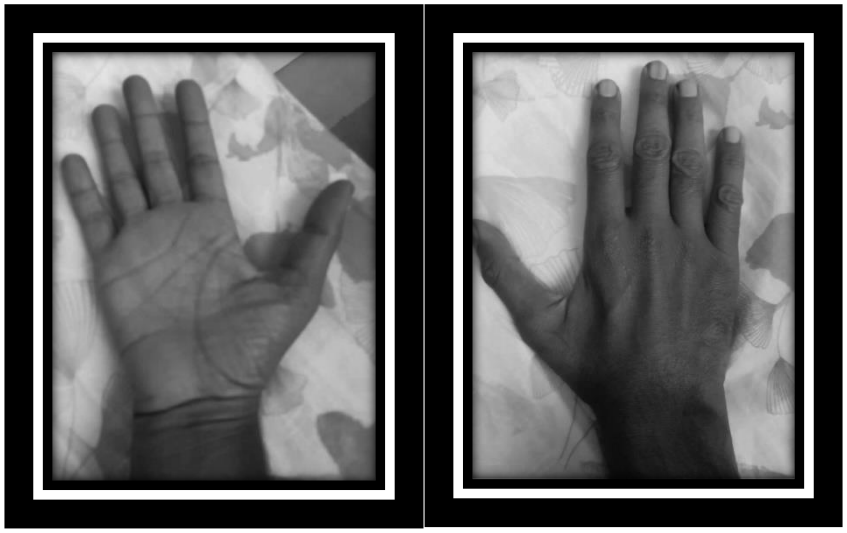

Figure 2 (Right Hand)- Clinical photograph showing wasting at the anatomical snuff box

On sensory assessment it was found that the superficial and deep sensations were intact, the reflexes were normal and the cranial nerves were intact. The tone of the muscles was normal, however, the muscle power of the hand and forearm muscles excluding brachioradialis was reduced.

Table 1- Manual Muscle Testing of Upper Limb Muscles

Muscles	Right	Left
Biceps Brachii	4	5
Triceps Brachii	4	5
Brachialis	4	5
Brachioradialis	4	5
Flexor Digitorum Superficialis	3-	4
Flexor Digitorum Profundus	3	4
Extensor Digitorum	3	4
Extensor Indicis	3	4
Opponens Pollicis	3	4
Flexor Policies Longus	3	4
Flexor Policies Brevis	3	4
Extensor Policies Longus	3	4
Extensor Policies Brevis	3	4
Abductor Policies Longus	3	4
Abductor Policies Brevis	3	4
Adductor Policies	3	4
Opponens Digiti Minimi	3	4
Abductor Digiti Minimi	2+	4
Extensor Digiti Minimi	3-	4
1,2,3,4, Lumbricals	3	4
1,2,3,4 Palmer Interossei	3	4
1,2,3 Dorsal Interossei	3	4

As noted in cases of HD, the brachioradialis muscle was spared. The balance and coordination were intact. On posture assessment, there was reduced cervical lordosis, the shoulders were protracted bilaterally, the right shoulder was depressed, the right scapula was abducted and there was decreased lumbar lordosis.

Diagnostic Assessment:

The MRI findings were found to be inconclusive since flexion/ extension films were not taken. NCV reports showed that the ulnar to median ratio (UN Ratio) (ADM/APB) was found to be 0.24 on the right and 0.44 on the left. The normal UN ratio is >0.6. A UN ratio of <0.6 is highly suggestive of HD.[12,13,14] It was noted in NCV reports that both sides were affected but the right side was more affected than the left side and the patient was asymptomatic on the left side. The disabilities of arm, shoulder, and hand (DASH) questionnaire was used to assess the upper limb function.

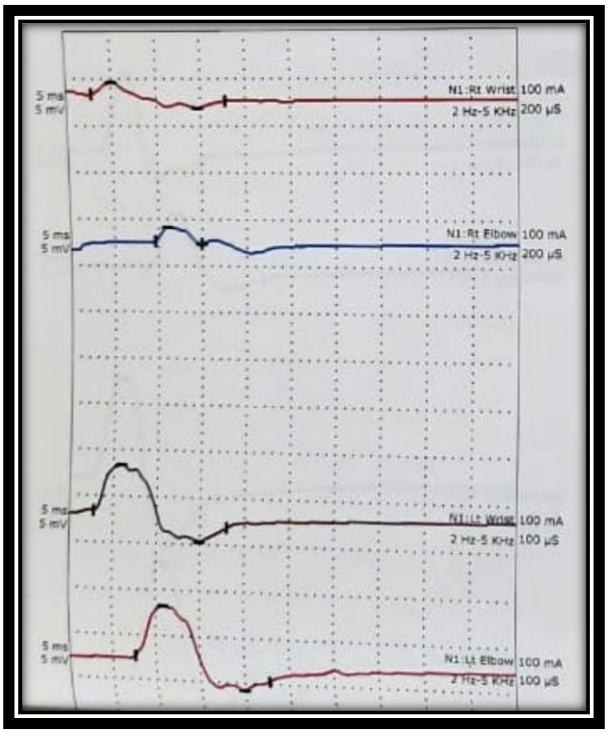

Fig 5: Motor Nerve Conduction Graph of Ulnar Nerve (ADM):

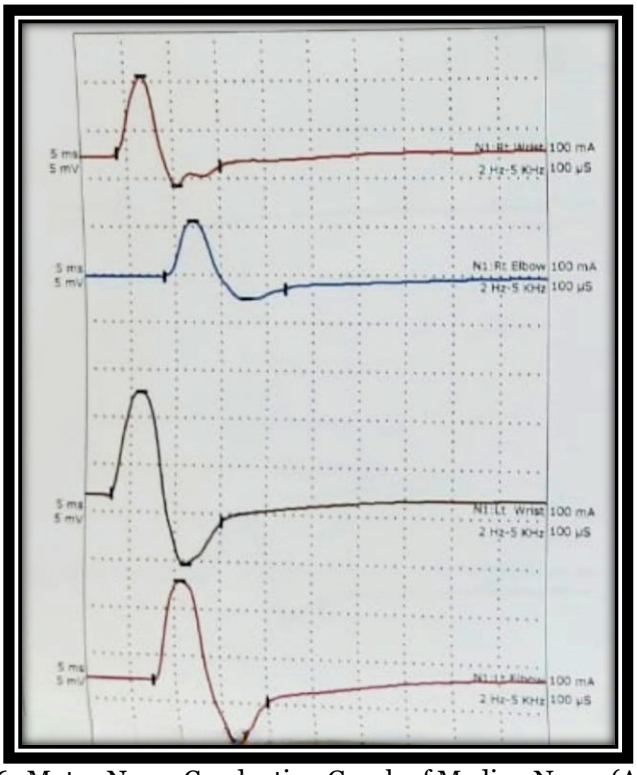

Fig 6: Motor Nerve Conduction Graph of Median Nerve (APB):

Therapeutic Intervention:

A physiotherapy protocol of 4 weeks was designed as shown in Table 2. For the first 2 weeks, the treatment was done under supervision 5 times/ week and the following 2 weeks treatment was given as a home program with a weekly follow-up. A cervical collar was prescribed since it controls neck flexion and thus limits the progression of the disease. Ergonomic advice was given which included taking a small height pillow or a towel roll and dropping the vision to do work rather than looking down to avoid neck flexion. Energy conservation techniques were advised. A post-treatment DASH was taken at the end of 4 weeks.

Table 2- Exercise Prescription (Supervised & Home Program)

Type of Exercise	Frequency of Exercise
Biceps curls with dumbbells	10 reps, 2 set
Triceps extensions with dumbbells	10 reps, 2 set
Wrist flexion, extension, radial deviation, and ulnar deviation with dumbbells	10 reps, 2 set
Hand gripping with gripper	10 reps, 2 set
Pinches with resistance springs	10 reps, 2 set
Thumb and all finger extensions with resistance	10 reps, 2 set
Scapular retractions	10 reps, 2 set
Cervical isometrics	10 reps, 10 seconds hold, 2 set
Transverse abdominis activation exercise	10 reps, 2 set
Gripping with therapeutic putty	10 reps, 2 set

Follow-up and Outcomes:

At baseline, the DASH score was 76/150 which was improved to 72/150 after 4 weeks of intervention. Though the total DASH score was not clinically significant, individual components showed improvement in activities like turning a key, pushing a heavy door, placing an object on a shelf above your head, and difficulty in sleeping due to pain indicating improvement in the functional status of the patient. There was also an improvement in the component of feeling less capable, less confident, or less useful on the DASH scale implying an improvement in the psychosocial domain. Thus, overall there was an improvement in the biopsychosocial aspect of the patient's condition.

Discussion:

One study found that there was a significant improvement in DASH score after 6 weeks of intervention consisting of isometric neck exercises, use of pressure biofeedback for deep cervical flexors, and upper limb resistance exercises.[20] A strong stable core is essential to provide good spinal alignment, and stability of the thoracolumbar spine, pelvis, hips, and distal limb. Increased cervical kyphosis leads to instability in cases of HD; coupled with weak cervical core musculature.[9] Another study described how improving postural parameters improved neurophysiology, sensorimotor control, and autonomic nervous system function in subjects with modifiable neurological disorders.[11] A recent study published in the Journal of Physiology also concluded that after 4 weeks of strength training, there was an increase in motor neuron output from the spinal cord to the muscle.[10] Our study in addition to neck isometric and deep cervical flexor activation exercises also included scapular strengthening and core strengthening exercises to improve spinal alignment and indirectly modulate neurophysiology in this case of HD. We did find an improvement in DASH score by 4 points which was not clinically significant.

Our study shows an improvement in strength and function in our patient with HD after a tailor-made treatment protocol with a focus on scapular alignment and spinal alignment was given. Because this is a case study and there are no standard treatment protocols for Hirayama's Disease, with due caution we can suggest that the focus should be on symptom management and spinal alignment in cases of HD.

Informed Consent:

A written informed consent was obtained from the patient before the treatment and included permissions related to data usage for scientific publications.

Ethical Clearance: Ethical clearance was taken from Institutional Review Board

Source of funding – self

Conflict of interest - Nil

References:

1. Lay S, Gudlavalleti A, Sharma S. Hirayama Disease [Internet]. StatPearls Publishing; 2023. Available from: https://www.ncbi.nlm.nih.gov/books/NBK499913

2. Bohara S, Garg K, Mishra S, Tandon V, Chandra PS, Kale SS. Impact of various cervical surgical interventions in patients with Hirayama's disease—a narrative review and meta-analysis. Neurosurg Rev. 2021 Apr 21;44(6):3229–47.
3. Hayden ME, Kim J, Zsuzsanna Arányi, Wolfe SW. Outcome of Tendon Transfer for Monomelic Amyotrophy (Hirayama Disease). J Hand Surg. 2023 Jan 1;48(1):90.e1–5.
4. Iacono S, Vincenzo Di Stefano, Gagliardo A, Cannella R, Virzì V, Pagano S, et al. Hirayama disease: Nosological classification and neuroimaging clues for diagnosis. J Neuroimaging. 2022 Apr 8;32(4):596–603.
5. Fukutake T. Hirayama Disease can be Caused by Loss of Attachment of the Cervical Posterior Dura to the Pedicle due to Immunological Abnormalities of the Dura and Posterior Ligaments: A New Hypothesis. Brain Nerve. 2020 Dec 1;72(12):1371–81. (Japanese).
6. Malanga GA, Aydin SM, Holder EK, Petrin Z. Functional Therapeutic, and Core Strengthening. Springer eBooks. 2016 Nov 30;185–214.
7. Wänman J, Persson PA, Bobinski L. Hirayama's disease associated with cervical deformity and spinal cord compression: a case report from Sweden. Acta Neurochir (Wien). 2024 Feb 10;166(1).
8. Tian Y, Xie L, Jiang J, Wang H. Why the patients with Hirayama disease have abnormal cervical sagittal alignment? A radiological measurement analysis of posterior cervical extensors. J Orthop Surg Res. 2022 Jan 15;17(1).
9. Li Z, Zhang W, Wu W, Wei C, Chen X, Lin J. Is there cervical spine muscle weakness in patients with Hirayama disease? A morphological study about cross-sectional areas of muscles on MRI. Eur Spine J. 2020 Jan 16;29(5):1022–8.
10. Dideriksen J, Del Vecchio A. Adaptations in motor unit properties underlying changes in recruitment, rate coding, and maximum force. J Neurophysiol. 2023 Jan 1;129(1):235–46.
11. Oakley PA, Moustafa IM, Harrison DE. The Influence of Sagittal Plane Spine Alignment on Neurophysiology and Sensorimotor Control Measures: Optimization of Function through Structural Correction. IntechOpen eBooks. 2021 Jul 14.
12. Kalita J, Kumar S, Neyaz Z. Split hand index and ulnar to median ratio in Hirayama disease and amyotrophic lateral sclerosis. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2017 Jun 15;18(7-8):598–603.
13. Philippe Corcia, Bede P, Pradat PF, Couratier P, Vucic S, de Carvalho M. Split-hand and split-limb phenomena in amyotrophic lateral sclerosis: pathophysiology, electrophysiology, and clinical manifestations. J Neurol Neurosurg Psychiatry. 2021 Jul 19;92(10):1126–30.
14. Ghasemi M, Alizadeh M, Basiri K, Ansari B, Tayebi Khorami R. Abductor Pollicis Brevis/Abductor Digiti Minimi Compound Muscle Action Potential Ratio as a Diagnostic Marker for Amyotrophic Lateral Sclerosis. Caspian J Neurol Sci. 2023 Jul 1;9(3):162–8.
15. Sikakulya FK, Kiyaka SM, Olasinde AA, Kiswezi A. Hirayama disease with proximal upper limb involvement in an adolescent female: A case report. Int J Surg Case Rep. 2022 Sep 1;98:107577–7.
16. Mishra P. Hirayama Disease: Will Physiotherapy Help? Int J Health Sci Res. 2019 Jan 1;9(10):220–4.
17. Sachar S, Sivarajan A, CJ V, Sachar S. A rare case of Hirayama disease. Int J Radiol Diagnostic Imaging. 2022 Oct 1;5(4):101–3.
18. Calisgan E, Canbay A, Talu B, Tecellioglu M, Sevimli R. HIRAYAMA DISEASE: AN UNUSUAL CASE REPORT. Ann Med Res. 2018 Jan 1;25(4):772–2.
19. Sapkota N. Effect of Strengthening Exercises and Muscle Stimulator in Hirayama Disease Patients Having Muscle Atrophy and Weakness: A Case Report. AJHS. 2023;3(1):5.
20. Oraon A, Patel L, Sarkar B. Physiotherapeutic management of hirayama disease: A case report. 2022 Jan 1;16(1):37–7.
21. Ivy CC, Smith SM, Materi MM. Upper extremity orthoses use in amyotrophic lateral sclerosis/motor neuron disease: three case reports. Hand (N Y). 2014 Dec 1;9(4):543–50.