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Case report

Expanding the clinical spectrum of Hirayama disease: a rare case of Hirayama disease with ulnar nerve palsy

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Abstract

Hirayama disease, also known as monomelic amyotrophy, is a rare, self-limiting condition that primarily affects young males. It leads to muscle atrophy and weakness, especially in the hands and forearms, which is often due to cervical spinal cord compression during neck flexion. Here, we report a case of a 41-year-old female from rural India who presented with progressive weakness in her left hand, resulting from ulnar nerve compression. The patient was unable to actively extend the metacarpophalangeal joint of her left hand, thus substantially impairing her hand function. Notably, she did not exhibit autonomic symptoms, which are commonly present in cervical myelopathy. This case highlights the importance of considering Hirayama disease in females, despite its male predominance, as well as the need for early diagnosis and targeted rehabilitation. Timely intervention is essential for managing functional impairments and improving patient outcomes.

Keywords: Chronic disease, Hirayama disease, monomelic amyotrophy, muscle atrophy, ulnar nerve compression rehabilitation.

Musculoskeletal health plays an important role in maintaining an individual's quality of life. Muscles can be affected by direct trauma, such as during contact sports, falls, or overuse. Injury to the muscle in these cases is a sudden event that leads to acute pain with the ability to heal completely within a few weeks to months. Such injuries to muscles, even if short-lived, still have a negative impact on an individual's independence. Muscle injuries in sports can also have a debilitating effect on the career of an athlete or reduce their work efficiency. Approximately 1.7 billion adults suffer from musculoskeletal conditions worldwide, which contribute to their burden on life.[1]

Hirayama disease (HD), also known as juvenile muscular atrophy of the distal upper extremities or monomelic amyotrophy, is a unique neurological disorder that was initially identified by Keizo Hirayama, a Japanese neurologist. HD is marked by the early onset of weakness and muscle wasting that affects one side of the hand and forearm. This condition specifically involves the muscles that are linked to the C7-T1 myotomes, whereas the brachioradialis muscles remain unaffected.[2]

HD is characterized by progressive loss of muscle girth and strength. The involvement is limited to the forearms and hands and may be unilateral or bilateral. This condition is progressive in nature for a period of 12–24 months, followed by a plateau phase and ending abruptly. Numerous theories, such as "contact pressure theory," "tight dural canal theory in flexion," and "growth imbalance between the vertebral column and dural canal," have been previously postulated to understand the pathogenesis.[3] Chronic ischemic changes because of limited dural laxity in the anterior horn cells of the cervical spine are the causative factor.[4]

Mild ulnar nerve compression at the elbow can be regarded as a clinical characteristic of HD. [2] Furthermore, clinical signs often present as (i) weakness and muscle wasting in the distal areas, mainly affecting the ulnar side of one arm or asymmetrically in both arms; (ii) gradual emergence and development over a span of 3–5 years, subsequently leading to stabilization of the condition

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or a relatively mild phase; (iii) unsteady, heavy shaking in the fingers of the affected hand(s); (iv) slight temporary aggravation of symptoms when the person is exposed to colder environmental conditions; and (v) lack of measurable sensory impairment. [5]

In 2020, research was conducted to establish a clinician-led guideline for the diagnosis and treatment of HD using a modified Delphi process. A total of six countries were involved in the expert panel of the Delphi process, and this provided a single direction for clinical practice. [6]

Case report

Patient information

A 41-year-old female receptionist, with no history of trauma to the head, neck, or spine, presented with difficulty in performing sophisticated work, such as tying knots, and the inability to completely extend the fingers of the left hand. She presented with a medical history of hyperthyroidism for eight years, which was treated with medication. She was also diagnosed with left ulnar nerve palsy 20 years ago, post-full-term normal delivery. Two years ago, the patient started noticing changes in her right hand near the base of the thumb, and similar changes were noticed a year ago in the left hand. The patient did not experience tingling or numbness. The condition developed without substantial disturbances in her profession, as there was no difficulty in writing. Biochemical analysis revealed serum urea and creatinine levels of 26.0 mg/dL and 0.7 mg/dL, respectively.

Clinical findings

On physical therapy assessment, the patient was conscious, cooperative, and oriented to time, place, and person. Moreover, bilateral atrophy of the muscles on the thenar eminence was observed, indicating progressive neurogenic muscle wasting, with asymmetrical involvement where the right hand appeared more affected in terms of tropical skin changes and muscle wasting. The thumb was adducted at the carpometacarpal joint **(Figure 1A),** which suggests weakness of the abductor pollicis brevis, a common finding in distal upper limb involvement in HD. In addition, flexion of the 3rd to 5th metacarpophalangeal (MCP) joints on the left hand **(Figure 1B)** indicates clawing because of intrinsic muscle weakness, often referred to as "oblique amyotrophy" or "monomelic amyotrophy." Upon examination, sensory loss on the palmar aspect of the left ring finger was present. Range of motion examination of the cervical spine, shoulder, and elbow revealed a full functional range for all movements, even though strength was reduced to 2+ on the left side compared to 3 on the right side for shoulder extensors for manual muscle testing. The active range of MCP joint extension was absent on the left side. The passive range exhibited a full normal end feeling. Strength on the right side for power and precision grips was reduced.

Diagnostic assessment

The magnetic resonance imaging (MRI) findings revealed straightening of the cervical spine **(Figure 2),** thus indicating a loss of normal cervical lordosis. Osteophytes were observed anteriorly and posteriorly at various levels **(Figure 2),** along with disk desiccation at several intervertebral disk levels **(Figure 3).** Notably, spinal cord atrophy was present at the C5–C6 level **(Figure 3),** which is characteristic of HD. There was no evidence of vascular engorgement of the epidural space or vascular plexus, and a small perineural cyst was identified at the C6–C7 level on the right side. Although the MRI findings support the diagnosis, the absence of nerve conduction and electromyography studies limits the confirmation of neurogenic involvement. These studies could have provided objective evidence of anterior horn cell dysfunction, thereby further reinforcing diagnostic confidence. Their omission may be due to resource limitations or clinical judgment, but their role in differentiating HD from other motor neuron disorders is well documented in the literature. The patient was diagnosed with HD, a rare form of monomelic amyotrophy, and was referred for conservative management in the physical therapy department.

Diagnosis

The diagnostic findings are in accordance with HD. A diagnosis of monomelic amyotrophy, which is HD in its progressing phase, was made, with the presence of ulnar nerve palsy as a comorbidity.

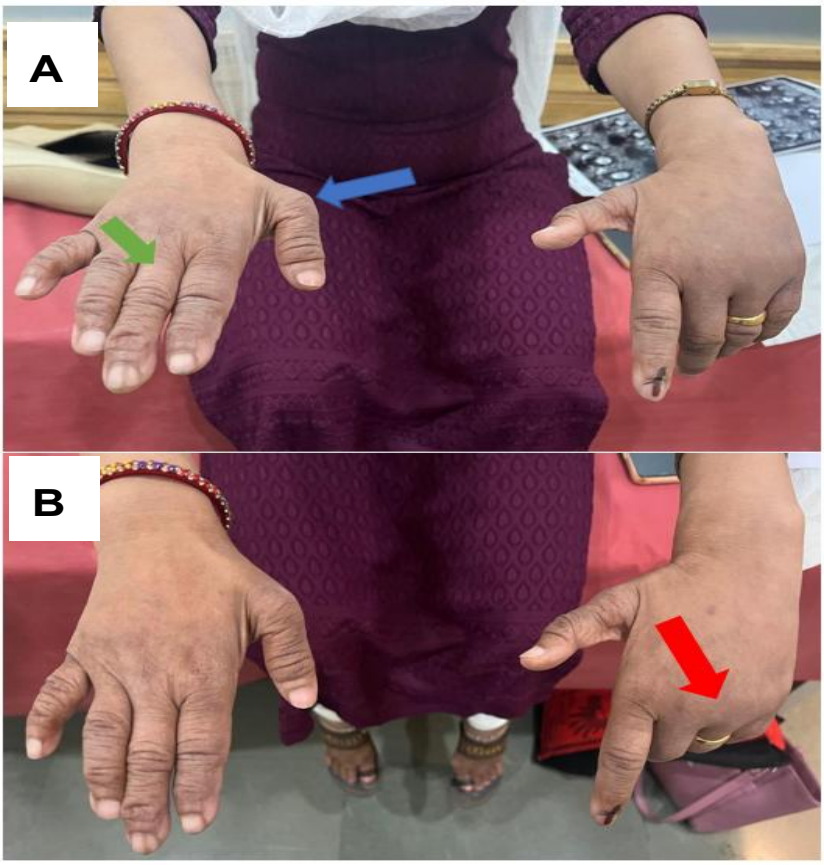

Figure 1. A) Superior view of the hand, blue arrow indicates the altered position of the right thumb. The green arrow indicates tropical skin changes; and **B)** The red arrow indicates the resting position of the left hand. The inability of the patient to keep the fingers in extension.

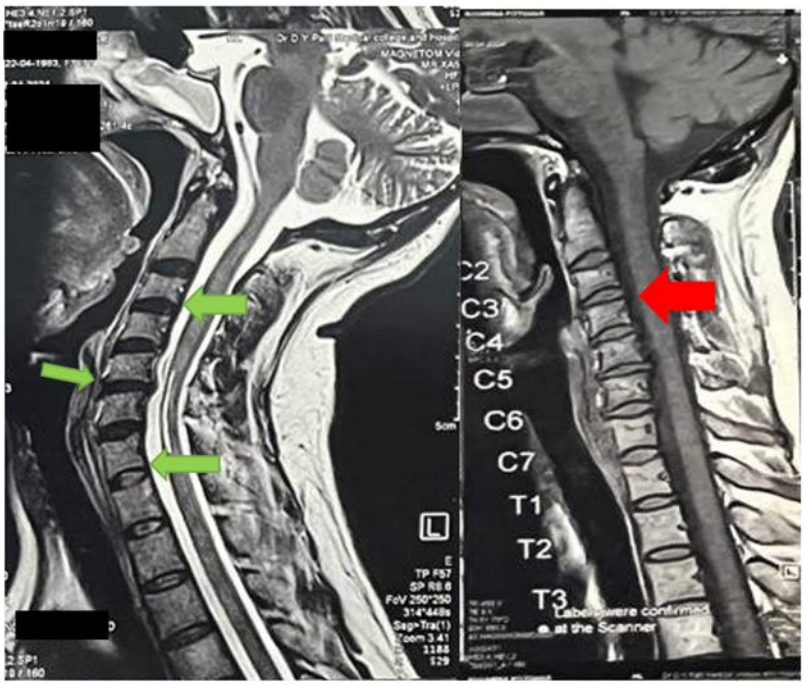

Figure 2. MRI of cervical spine in extension (left image), the arrow in green indicates osteophytes at multiple levels. The image on the right is an MRI in the neutral position of the cervical spine. The red arrow indicates in this image the loss of cervical lordosis. Disc bulge from C4-C7 is seen, intending the anterior thecal sac.

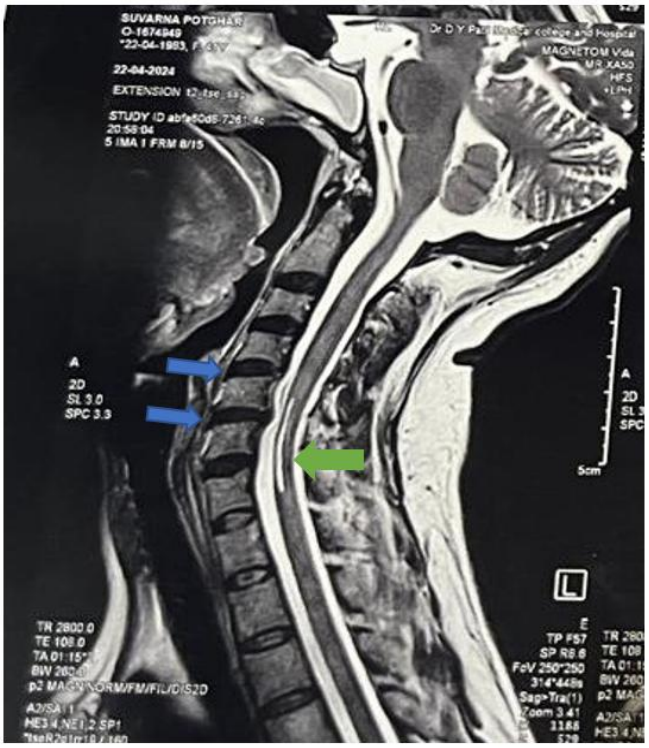

Figure 3. MRI of cervical spine in extension. The blue arrow indicates intervertebral disc desiccation. The green arrow is indicative of spinal cord atrophy.

Treatment

During physiotherapy treatment, the patient continued her prescribed medication for hyperthyroidism, including 15 mg/day of methimazole and multivitamins. A comprehensive rehabilitation protocol was designed to address the muscle weakness, joint integrity, and functional deficits associated with HD. Active range of motion exercises for the cervical spine, shoulder, elbow, and wrist were prescribed three times a day with 12–15 repetitions to maintain joint mobility and prevent stiffness. Moreover, considering the presence of flexion deformity, passive stretching of the left MCP joint using the right hand was introduced to maintain flexibility and prevent contracture. Muscle-strengthening exercises were incorporated using a theratube positioned just proximal to the wrist joint to enhance the activation of the elbow and shoulder flexors, extensors, and scapular stabilizers, with two sets of 15 repetitions. This approach aimed to counteract the progressive muscle atrophy while improving upper limb function. Grip strengthening was emphasized using a stress ball, where the patient was required to hold and release the object with controlled effort, mirroring the repetition scheme of other exercises to enhance endurance and motor control.

Fine motor training included finger-tapping exercises, where the patient was instructed to spread the fingers and tap them against the thumb in a controlled yet rapid manner. This targeted the improved coordination and independent finger movement, which are often compromised in patients with HD. Finger extension exercises using a rubber band around the fingers and thumb helped facilitate extensor muscle activation and prevent flexion contractures because of muscle imbalance. In addition, therapeutic putty exercises were introduced, starting with soft putty (85g) and progressing through a series of grip patterns, including cylindrical grip, hook grip, and isolated fingertip squeezing. This progressive approach aims to restore functional hand strength and dexterity, which are essential for daily activities and fine motor skills.

Discussion

HD is a rare condition, with very few published case reports and clinical images.[7-9] It is a rare neurological condition with weakness that is asymmetrical and atrophy of the hand muscles. Autonomic involvement is very rare, with unusual features, as reported by Anuradha S, *et al*.[10] In comparison to previously reported cases of HD, our patient represents a unique case because of the onset of symptoms in the fourth decade of life. Most cases are reported in adolescents or young adults, with a peak incidence in the second to third decades of life. In contrast to the typical age range of 15–35 years, the onset during the fourth

decade of life in our patient suggests that the disease may have a broader onset age range than traditionally believed. In addition, HD predominantly affects young men, with an increased prevalence in countries such as Japan and India. Although rare worldwide, the condition has been increasingly recognized in other regions as well. Our case emphasizes the necessity of global awareness as cases are being recognized outside this region. It predominantly affects the lower cervical spine, and these findings are parallel to the findings of our case study. Moreover, alteration in sensations, reflexes, and cerebellar deficits is a very rare finding, and clinical features may include tremors that increase with cold.[11] Multiple studies state the seven features of the criteria required for the diagnosis of HD.[9, 12]

The exact pathogenesis of HD is unknown, but cervical myelopathy with neck flexion is the most accepted hypothesis.[13] During neck flexion, the cervical cord moves forward, which results in the compression of the anterior spinal artery, thus leading to ischemia in the anterior horn cells, particularly in the C5–C7 region. This ischemic damage is responsible for the muscle weakness and atrophy observed in the affected limb, especially in the distal upper extremities. Genetic predispositions and anatomical variations play key roles in the development of HD. Spinal cord displacement may be exacerbated by abnormal cervical spine mobility and may contribute to the pathophysiology of the disease. Unlike the other reported studies, our patient was in the fourth decade of life when her symptoms started to precipitate. Even though the condition is self-limiting, this patient also presented with ulnar nerve palsy, which was acquired at the time of post-full-term normal delivery. Ulnar nerve compression during pregnancy is known as Guyon's tunnel syndrome.[14] The presence of this adds to the burden on the patient's quality of life. Furthermore, HD must be differentiated from other conditions that cause asymmetric upper limb weakness and atrophy. The differential diagnosis includes cervical spondylotic myelopathy, amyotrophic lateral sclerosis, and multifocal motor neuropathy. Cervical spondylotic myelopathy is a degenerative condition where the compression of the spinal cord can lead to similar symptoms, such as muscle weakness and atrophy, particularly in the hands. However, unlike HD, cervical spondylotic myelopathy typically presents with a gradual, progressive course and is not usually self-limiting.

The cooccurrence of ulnar nerve palsy in our patient adds to the complexity of her rehabilitation process. Ulnar nerve compression, particularly during pregnancy, is a well-documented phenomenon that is often due to compression at Guyon's canal. This compression can lead to considerable motor and sensory deficits in the hand, affecting the ability to perform fine motor tasks, such as gripping or holding objects. Guyon's tunnel syndrome is commonly observed in pregnant women because of increased fluid retention, hormonal changes, and postural shifts, which may lead to nerve compression. When combined with the muscle weakness and atrophy caused by HD, the patient's quality of life was further compromised. Inability to perform activities of daily living that involve tying hair, holding objects, eating food, riding a bike, and other household chores, should the atrophy progress to a greater extent before cessation of the condition, can lead to a negative impact on the patient's perspective of life. Our aim through rehabilitation was to maintain the patient's current level of activity and improve certain aspects of grip by constantly recruiting the muscle, as well as reeducating. Furthermore, we aimed to alter certain movement patterns to teach the patient the ability to perform activities on her own. As the patient was staying far from the outpatient department physiotherapy facility, follow-up and regular progression could not be maintained. Further research on effective rehabilitative exercises for HD, patients' psychology, and the abrupt cessation of the disease is required.

A graphical abstract summarizing the clinical and rehabilitative aspects is shown in **Figure 4.**

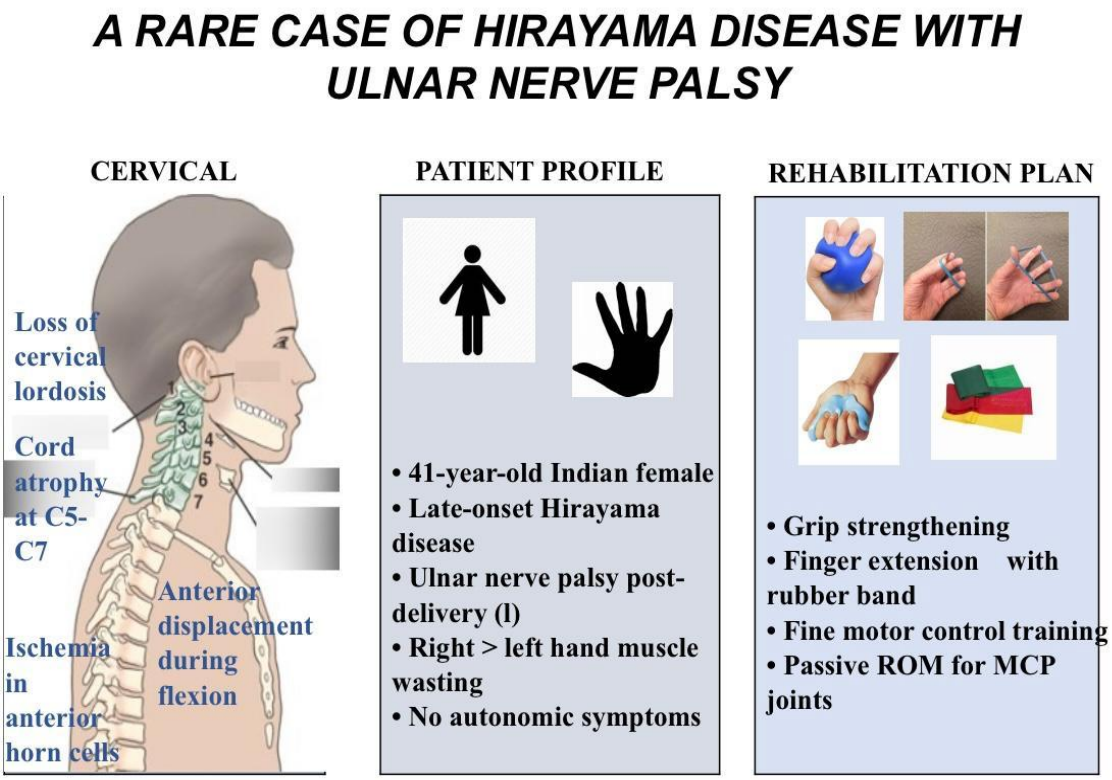

Figure 4. Graphical abstract summarizing the clinical presentation, diagnostic features, and rehabilitation approach for a 41-year-old female with Hirayama disease and ulnar nerve palsy.

Conclusion

This case report highlights the role of early diagnosis and structured physiotherapy in managing HD, with a specific focus on maintaining joint mobility, muscle strength, and functional independence. A tailored rehabilitation approach, combined with patient education, can help slow disease progression and improve the patient's overall quality of life. Furthermore, emphasizing adherence to therapy and long-term monitoring is essential in mitigating functional decline and optimizing patient outcomes.

Author contributions

CK and MJ conceptualized the design of the work, CK and SM done the analysis and interpretation of data for the work. CK, SM and MM have done the Drafting the work or revising it critically for important intellectual content. All four authors have approved of the version to be published. All the authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of interest statement

All authors have completed and submitted the International Committee of Medical Journal Editors Uniform Disclosure Form for Potential Conflicts of Interest. All authors declare that they have no conflicts of interest.

Data sharing statement

Data generated or analyzed for the present report are included in this published article. Further details are available from the corresponding author on reasonable request after deidentification of the patient whose data are included in the report.

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