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A CASE STUDY ON A COMPREHENSIVE REVIEW OF HIRAYAMA DISEASE

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ABSTRACT

Adolescent boys are the main victims of Hirayama disease (HD), a rare, self-limiting cervical myelopathy that manifests as gradual, asymmetric weakening and atrophy of the distal upper limb. The condition is caused by anterior displacement of the posterior dura mater, which compresses the spinal cord dynamically during neck flexion. HD stabilises following an initial active phase, in contrast to progressive motor neurone disorders. Flexion MRI, which shows distinctive dural changes and venous congestion, is the key to the diagnosis. Permanent spinal cord injury can be avoided with early cervical immobilisation treatments. Surgical procedures such as anterior cervical decompression and fusion may be useful in situations that are progressing or refractory. The pathophysiology, clinical presentation, imaging, differential diagnosis, and therapeutic approaches of HD are all summarised in this study. For prompt diagnosis and better outcomes, greater worldwide awareness and sophisticated imaging are crucial.

KEYWORDS: cervical myelopathy, flexion MRI, motor neuron disorder, distal upper limb weakness, dynamic spinal cord compression, adolescent males, cervical collar.

INTRODUCTION

First identified by Keizo Hirayama in 1959, Hirayama disease (HD) is a rare neurological condition that primarily affects male adolescents and young adults. The distal upper limb

is marked by gradual, asymmetric muscular wasting and weakness, especially in the hand and forearm muscles. This condition is often referred to as monomelic amyotrophy. Hirayama illness is self-limiting and does not cause sensory loss or upper motor neurone symptoms, in contrast to traditional motor neurone diseases.[1] The primary mechanical cause of HD is thought to be the forward displacement of the posterior dura mater during neck flexion, which causes the cervical spinal cord to be dynamically compressed and the anterior horn cells to sustain persistent ischaemic damage. Because of this distinct pathophysiology, dynamic magnetic resonance imaging (MRI), particularly while the neck is flexed, is crucial for a precise diagnosis. Hirayama disease is becoming more well-known globally, despite being more common in Asian nations like China, India, and Japan. However, because of its unusual presentation and rarity, it is still underdiagnosed. Since prompt action can stop the progression of the disease and avoid chronic disability, early recognition is essential. The goal of this study is to compile the most recent information on Hirayama disease's clinical manifestation, imaging characteristics, diagnostic methodology, aetiology, and potential treatment options. We intend to increase clinician awareness and enhance early detection and care of impacted persons by combining ideas from the body of existing work.[2,3]

2. History

The majority of instances of Hirayama disease (HD), a rare, non-progressive motor neurone condition, have been documented from Asian nations, namely China, Taiwan, Japan, and India. But as awareness has grown and imaging technology has improved, cases have been found all across the world, even in Western nations, raising the possibility that the illness is underdiagnosed rather than genuinely uncommon. With a male-to-female ratio of roughly 20:1, the illness primarily affects young males, however, there have been a few documented cases in girls. It usually manifests between the ages of 15 and 25, which corresponds to the teenage growth spurt. Because of this temporal association, there is conjecture that the mechanical pathogenesis seen in HD may be exacerbated by rapid growth of the vertebral column beyond the expansion of the dural sac. Because of the disease's rarity and diagnostic difficulties, prevalence data are scarce. Nonetheless, some information has been gleaned from modest epidemiological studies conducted in high-prevalence areas: One regional research in Japan found the incidence to be 1.4 per 100,000 males. According to Indian research, 5–10% of all motor neurone disease cases in neurology clinics may have HD. Awareness campaigns and routine

cervical flexion MRI use have greatly enhanced the detection rate in China and Taiwan. A consistent genetic or familial pattern has been found, supporting the notion that HD is primarily sporadic. Though the evidence is still equivocal, some theories point to a potential hereditary vulnerability in particular communities or families. In predisposed people, environmental or mechanical variables such repetitive neck motion during physical activities, bad posture when studying or working, or habitual neck flexion may also play a role in the onset or worsening of the condition.[4,5]

3. Pathophysiology and Etiology

Among motor neurone illnesses, Hirayama disease (HD) has a distinct pathogenesis that is typified by a dynamic mechanical compression of the lower cervical spinal cord, especially during neck flexion. In contrast to other neurodegenerative diseases, HD is brought on by extrinsic vascular and mechanical causes that result in localized ischaemia rather than intrinsic neuronal loss.

3.1 The Hypothesis of Mechanical Compression

According to the most frequently recognised idea, the posterior dural sac shifts anteriorly during neck flexion because it is not adequately attached. Particularly at C5 to C7 levels, this aberrant movement compresses the cervical spinal cord and narrows the spinal canal. The problem frequently eludes diagnosis on routine MRI because this flexion-induced cord compression does not occur in the neutral position. Repeated microtrauma caused by bending over time results in

- Wasting and denervation of muscles innervated by the impacted cervical segments.
- Ischaemia of the anterior horn cells as a result of impaired blood flow in the anterior spinal artery area; Flexion MRI, which shows forward displacement of the posterior dura and enlargement of the posterior epidural space, frequently filled with a congested venous plexus, is the best way to illustrate this dynamic mechanism.[6]

3.2 Growth Imbalance Hypothesis

During adolescence, a time of significant physical growth, HD frequently appears. Increased slack in the dura may result from unequal growth between the dural sac and the spinal column, according to one idea. The spinal cord is more vulnerable to compression because of this anatomical mismatch, which allows the dura to move excessively forward during flexion.

3.3 Vascular Insufficiency

The anterior horn cells may sustain repeated ischaemic injury as a result of chronic flexion-induced compression that disrupts venous outflow and artery supply. A vascular ischaemic mechanism has been supported by autopsy and histological investigations that have revealed localised necrosis and gliosis restricted to the cervical spinal cord's anterior horns.[7]

3.4 Role of Connective Tissue Laxity

Congenital or acquired laxity of the posterior dural attachments may be a significant factor, according to some researchers. In people with hypermobility or connective tissue problems, insufficient tethering may make dural movement during neck flexion worse.

3.5 Genetic and Environmental Factors

The majority of HD cases are sporadic, and no consistent genetic alterations have been connected to the condition to yet. Nonetheless, in rare cases, familial clustering has been documented, pointing to a potential hereditary susceptibility. Genetically predisposed people may experience external stressors from environmental triggers such as repetitive neck flexion activities, sustained poor posture (e.g., students studying for long hours), or specific sport.[8]

4. Clinical Features

When combined with the right imaging, Hirayama disease (HD) can be distinguished from other motor neurone illnesses and myelopathies thanks to its unique clinical profile. Early detection of these characteristics is essential for timely diagnosis and treatment.

4.1 Age of Onset and Progression

Male adolescents between the ages of 15 and 25 are usually affected by HD. Over the course of one to five years, the illness typically starts slowly and advances before plateauing or stabilising. One important characteristic that sets it apart from progressive neurodegenerative diseases like amyotrophic lateral sclerosis (ALS) is its self-limiting nature.

4.2 Limb Involvement

Unilateral or asymmetric weakness and wasting of the distal upper limb, especially the hand and forearm muscles (C7–T1 myotomes), are the hallmarks of HD. Perhaps as a

result of hand dominance and increased use, right-sided participation is more prevalent. Weakness typically stays asymmetrical, but it can occasionally become bilateral.[9]

4.3 Pattern of Muscle Atrophy

The following muscles are impacted:

- the thenar and hypothenar muscle.
- The first interosseous muscles dorsally.
- The forearm's flexor and extensor muscles.
- The "oblique amyotrophy" pattern, which is thought to be pathognomonic, results from the disorder sparing the brachioradialis.

4.4 Sensory and Reflex Findings

- There is no evidence of sensory loss.
- Although they may be somewhat diminished in the affected limb, deep tendon reflexes are typically maintained.
- Absence of upper motor neurone symptoms, such as spasticity, hyperreflexia, or the Babinski sign, which are used to differentiate HD from ALS or cervical spondylotic myelopathy.[10]

4.5 Cold Paresis

- Known as cold paresis, many patients report that their symptoms worsen in cold situations.
- This condition might show itself as greater hand weakness or clumsiness when exposed to cold temperatures.

4.6 Absence of Bulbar or Lower Limb Involvement

- HD differs from other neurodegenerative diseases in that it does not impact lower limbs, respiratory function, or bulbar muscles.
- Additionally, cognitive and sensory abilities are unaffected.

4.7 Disease Course

HD's normal course is monophasic

Phase of progression: usually lasts six months to five years.

Phase of stability: symptoms level off and don't become worse.

Disability can be prevented or reduced with early intervention during the progressive

period.[11]

5. CASE REPORT

5.1 Patient Presentation

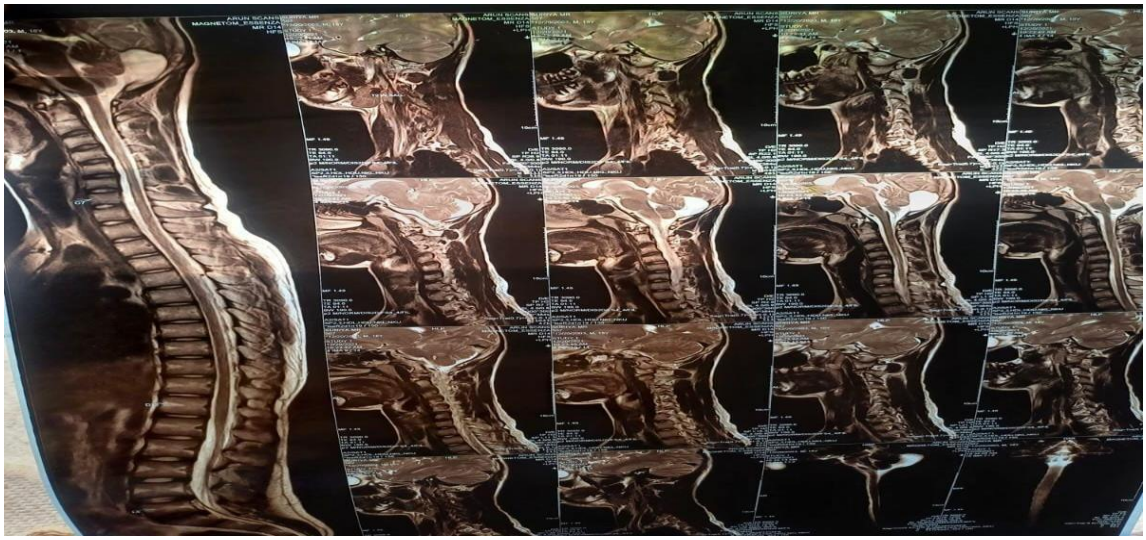
A left-handed male patient, age 21, has been experiencing growing weakness and thinning of his left hand and forearm for the past five years. No numbness, discomfort, or trauma history was present. The patient denied having any trouble eating, writing, dressing, etc. There was no contribution from family history.

5.2 Clinical Examination

- A neurological examination showed that the right thenar, hypothenar, and interosseous muscles were markedly weak and wasted.
- Mild weakness in the left wrist and finger extensors (MRC grade 4/5).
- Deep tendon reflexes were maintained, no sensory loss or fasciculations observed.
- The cranial nerves and lower limbs were both in normal condition.
- There was evidence of cold paresis, a condition where symptoms became worse when exposed to cold.
- There is little weakness in the right upper limb.

5.3 Investigations

- Without any sensory involvement, the EMG revealed persistent denervation alterations in the left side's C7, C8, and T1 myotomes.
- Mild cervical kypotic deformity at the C5 level on a cervical spine MRI in the neutral posture.

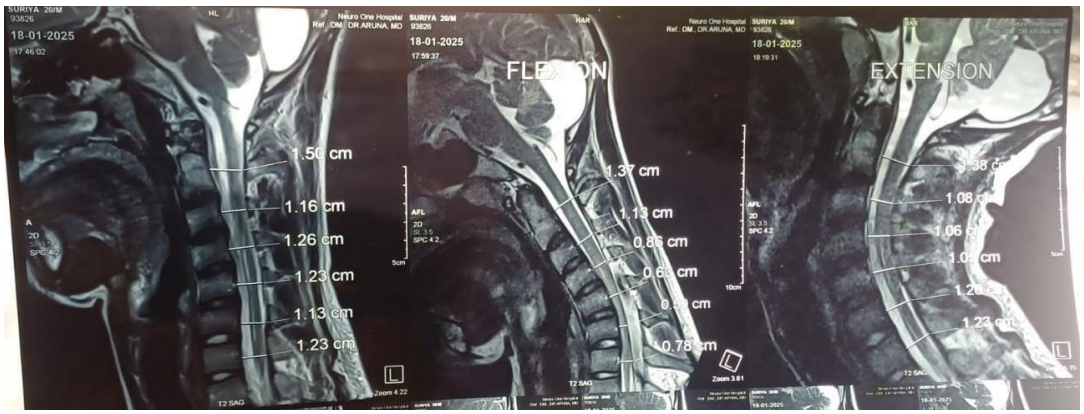


MRI Image of Neutral Position

MRI of cervical spine- Flexion-Extension Dynamics (Key Feature in Hirayama Disease)

Flexion pictures showed

- The posterior dura migrated forward, which is a hallmark of Hirayama illness.
- Considerable cord compression between C5 and C7, with noticeable displacement and constriction of the chord during flexion.
- Epidural venous engorgement—a dynamic change during neck flexion.



MRI image of flexion and extension position

Management and outcome

The patient received a hard cervical collar to wear all day and was counselled against excessive neck flexion. Additionally, he began physical treatment to strengthen his hands. After a year of monitoring, his symptoms levelled off and only gradually worsened. He returned to his studies after making ergonomic and posture corrections.

6. Diagnostic Evaluation

A high index of clinical suspicion and the use of flexion cervical MRI, which is essential for visualising the dynamic alterations causing the disease, are essential for the accurate and prompt diagnosis of Hirayama disease (HD). Without specialised imaging and electrodiagnostic testing, routine neurological tests could not yield a definitive result.

6.1 Clinical Diagnosis

Considering the results of the physical examination and the distinctive history:

- Self-limiting illness history.
- Slowly progressive, unilateral or asymmetric weakness and atrophy of the hand and forearm.
- No evidence of upper motor neuron dysfunction or sensory loss.[12]

6.2 Electrophysiological Studies

The diagnosis is supported by electromyography (EMG) and nerve conduction studies (NCS) because they show

- Normal sensory nerve action potentials, which help distinguish HD from peripheral neuropathies.
- Chronic denervation and reinnervation in C7, C8, and T1 myotomes.
- No indication of widespread anterior horn cell involvement, as in ALS.[13]

6.3 Magnetic Resonance Imaging (MRI)

- When done in both neutral and flexion positions, MRI is the gold standard for diagnosing HD.

Findings in the neutral position could consist of

- Localised atrophy of the lower cervical spinal cord (C5–C7).
- The cord flattens asymmetrically; the posterior dural sac and lamina no longer join.
- Key characteristics of the flexed position include

- Anterior displacement of the posterior dura mater.
- A crowded venous plexus fills the enlarged posterior epidural space.
- A distinctive crescent-shaped enhancing lesion or flow void on post-contrast T1 or T2- weighted images.
- Spinal cord compression, frequently without intervertebral disc herniation. These results validate that the disease is dynamic and reversible.[14]

6.4 Other Imaging Modalities

When MRI is unavailable or inconclusive, **dynamic CT myelography** is occasionally utilised; however, it is invasive.

Cine and ultrasound MRI: New methods for evaluating spinal cord dynamics and dural movement during flexion.

6.5 Diagnostic Criteria

The following are typical recommended diagnostic criteria (taken from the literature)

1. The distal upper limb weakness and atrophy in a young guy
2. No signs of higher motor neurons or sensory neurons
3. EMG evidence of anterior horn cell involvement
4. Dynamic cervical MRI showing forward displacement of the posterior dura during flexion.

7. Differential Diagnosis

Given its early onset, localised muscle atrophy, and mild onset, Hirayama disease (HD) might be mistaken for a number of distinct neurological conditions. Since the prognosis and treatment plan differ greatly, it's imperative to accurately distinguish HD from them.

7.1 Amyotrophic Lateral Sclerosis (ALS)

Key differences

- ALS usually manifests after the age of forty.
- Usually accompanied by bulbar symptoms, extensive muscle fasciculations, and a progressive course, this condition involves both upper and lower motor neuron indications.
- In ALS, the EMG demonstrates extensive anterior horn cell involvement.[15]

7.2 Cervical Spondylotic Myelopathy (CSM)

exhibits the following symptoms

- Sensory anomalies, hyperreflexia, and spasticity.
- Imaging demonstrates ligament hypertrophy, osteophytes, or disc degeneration resulting in spinal cord compression.
- Symptoms are not posture-dependent like HD is.

7.3 Multifocal Motor Neuropathy (MMN)

- This asymmetric distal limb weakness is caused by immune-mediated neuropathy.
- Although it can resemble HD, MGN usually entails conduction block on NCS; it responds to IVIG therapy; and its sensory nerves are maintained, though they may exhibit minor anomalies.

7.4 Spinal Muscular Atrophy (SMA)

Genetic testing (such as SMN1 deletion) is used to diagnose genetic disorders, which typically manifest in infancy or youth and cause symmetrical weakness, frequently involving proximal muscles.[16]

7.5 Post-Polio Syndrome

- Found in patients with a history of poliomyelitis;
- Symptoms include fatigue and weakness that develop gradually.
- History and serology aid in distinguishing it from HD.

7.6 Brachial Plexopathy or Cervical Radiculopathy

- Frequently linked to paresthesia, discomfort, or sensory loss.
- Unlike HD, EMG/NCS demonstrates sensory involvement.
- MRI may reveal anomalies in the plexus or nerve root compression.

8. Management Strategies

Preventing irreversible spinal cord injury and stopping the disease's progression during its active phase are the main objectives of Hirayama disease (HD) management. Early and adequate care, especially during the progressive phase, can greatly enhance long-term outcomes because the disease frequently has a self-limiting course. The two main categories of management are conservative and surgical.

8.1 Conservative Treatment

8.1.1 Cervical Collar Immobilisation

- The cornerstone of care, particularly in the progressive stage (often the first one to five years)
- To reduce dynamic spinal cord compression, a tight cervical collar is recommended to restrict neck flexion.
- Depending on symptom stabilization, a 12- to 24-month period is advised.
- Research indicates that wearing a collar early on can slow the progression of the illness and, in certain situations, permit a partial recovery.[17]

8.1.2 Physical Therapy

- Incorporates range-of-motion exercises and strengthening of unaffected muscles.
- Strives to preserve muscular function and prevent contractures.
- Strongly advises against repetitive neck flexion and heavy lifting activities.

8.1.3 Observation

Close observation may be necessary for patients with very modest symptoms or those in a stable (plateaued) phase. MRI tests and routine neurological examinations can be used to monitor disease activity.

8.2 Surgical Intervention

Surgery is often saved for situations that worsen or don't improve with conservative treatment.

Some indications are

Despite wearing a collar, the weakness persisted; a flexion MRI revealed severe spinal cord compression; and the conservative treatment was not well followed.[18]

8.2.1 Anterior Cervical Decompression and Fusion (ACDF)

Decompressing the spinal cord and fusing the impacted levels is the most often done technique to stop dynamic movement. It can successfully stop more neurological damage.

8.2.2 Dural Tacking or Posterior Cervical Duraplasty

Makes an effort to secure the posterior dura to the spinal canal in order to stop anterior

shift during flexion. With or without a laminectomy, it can be performed. Many centers still view it as an auxiliary or experimental treatment.

8.2.3 Posterior Instrumentation and Fusion

Tries to prevent anterior shift during flexion by securing the posterior dura to the spinal canal. It can be done with or without a laminectomy. It is still considered an experimental or supplemental treatment in many locations.

8.3 Prognosis and Long-Term Management

- Most patients stabilize after the initial progressive phase. With early intervention (collar use), many experience no further deterioration.
- However, muscle bulk and strength may not fully recover, especially if the diagnosis is delayed.

Long-term follow-up is essential to monitor

- Functional status
- MRI changes
- Patient compliance

9. Recent Advances and Future Directions

Even though Hirayama disease (HD) has been known for a number of decades, new developments in imaging, surgery, and a better comprehension of its pathophysiology are changing how the condition is diagnosed and treated. To improve diagnostic standards, find early biomarkers, and assess long-term therapy results, further research is necessary.[19,20]

Advanced Imaging Techniques

9.1.1 Cine MRI and Dynamic Studies

- By verifying dynamic cord compression, cine MRI helps distinguish HD from other compressive myelopathies.
- It allows real-time visualization of cerebrospinal fluid (CSF) flow and dural movement during neck flexion.
- When routine flexion MRI is inconclusive, it may enable early diagnosis in borderline or unusual instances.

9.1.2 Diffusion Tensor Imaging (DTI)

- DTI studies have revealed microstructural cord alterations even in regions that appear normal on conventional MRI.
- DTI is a sort of advanced MRI that evaluates the integrity of the white matter tract.
- could be applied to track therapy response and identify subclinical progression.

9.2 Electrophysiological Advances

- To more accurately measure motor neuron loss, researchers are investigating the use of quantitative EMG and motor unit number estimation (MUNE).
- These could provide a non-invasive way to assess post-treatment recovery and disease activity.

9.3 Biomarker Research

The following biomarkers are being studied despite the lack of specific biomarkers for HD

- Serum neurofilament light chain levels
- Inflammatory cytokines
- Genetic or connective tissue markers (in cases that may be familial)
- Biomarkers may aid in risk stratification or the identification of patients who may have a more aggressive course.

9.4 Surgical Innovation

- Evaluations are being conducted on minimally invasive methods for anterior fusion and duraplasty.
- In cases of complex or multilayer disease, intraoperative navigation and customized 3D-printed spinal implants are being investigated.

9.5 Artificial Intelligence (AI) and Machine Learning

To automatically identify dural displacement and other MRI characteristics of HD, artificial intelligence techniques are being developed. could help in early detection, particularly in facilities with little experience in radiology.

9.6 Awareness and Global Reporting

- Since underdiagnosis is still prevalent outside of Asia, recent initiatives have concentrated on increasing awareness of HD.

Multicenter research and global registries are required to more precisely define:

- Natural history
- Treatment response
- Environmental or genetic factors.

9.7 Foot Neuropathy Treatment

Although Hirayama illness mostly affects the upper limbs and cervical spinal cord, new developments in the management of peripheral neuropathies, especially those affecting the lower extremities, may provide clues for more comprehensive neuroprotective and regenerative approaches.[21]

Emerging treatments for foot neuropathy, such as diabetic or compressive neuropathies, include

- Neuromodulation methods (such as spinal cord stimulation and transcutaneous electrical nerve stimulation [TENS]).
- Neurotrophic agents (such as alpha-lipoic acid, B vitamins, and nerve growth factor analogues)
- Stem cell therapy and extracellular vesicle treatments, which are being studied for their potential to promote peripheral nerve regeneration
- Laser therapy exhibits promise in enhancing microcirculation and nerve conduction.
- The use of gene therapy, especially for hereditary neuropathies such as Charcot-Marie-Tooth illness.

These treatments may have translational potential for anterior horn cell preservation or regeneration in Hirayama disease, despite the fact that they have mainly been investigated in peripheral neuropathies of the lower limbs. Future neuroprotective techniques targeted at reducing flexion-induced ischemia in the cervical cord may be guided by an understanding of the molecular and vascular pathways driving neuropathic damage in the foot.

10. CONCLUSION

Young men are the main victims of Hirayama disease, an uncommon, self-limiting cervical myelopathy that manifests as atrophy and weakening of the distal upper limb muscles that develops gradually. It differs from other motor neuron and myelopathic

illnesses due to its distinct etiology, which involves dynamic spinal cord compression during neck flexion. Due to a lack of knowledge and the requirement for specialized imaging, the disease is still underdiagnosed despite its distinctive clinical and radiological symptoms, particularly outside of Asia. To avoid irreparable neurological impairments, early detection and diagnosis are essential, mostly via flexion MRI. While surgical intervention may be necessary in certain advanced instances, conservative care with cervical immobilization continues to be the cornerstone of treatment during the active period of the disease. Developments in neurology, imaging, and surgery keep improving our knowledge of and strategy for treating this illness. To determine the actual prevalence, optimize treatment plans, and enhance long-term results for individuals with Hirayama disease, multicenter studies, standardized diagnostic procedures, and increased global awareness is crucial.

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