Hirayama Disease

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Introduction

Hirayama disease, is an uncommon, slowly progressive disease that affects mostly men in their second to third decade of life.[1] It was first reported in 1959 by Hirayama in Japan[2] · It has an insidious onset with atrophy occurring primarily unilaterally in the distal upper extremities.[1] Typically the brachioradialis muscle is spared.[1] . Hirayama disease is believed to be caused by chronic micro-vascular ischemic damage to the lower cervical spinal cord that occurs during neck flexion.[2]

Case 1

A 23-year-old Hispanic male presented with progressive weakness of upper and lower extremities. He reported a six years history of progressive muscle weakness in his upper limbs and weakness to strengthening exercise. He was given birth after an uneventful pregnancy and never had any alterations in his developmental milestones. Past surgical history is positive for presumptive tendon transposition due to weakness of left upper extremity at age 8.

Physical exam revealed atrophy of left shoulder girdle (Figure 1A), asymmetric atrophy of third and fourth left dorsal interousseous muscles (Figure 1B) with preservation of the brachioradialis consistent with oblique atrophy (Figure1C).

An Electromyography test was ordered for further evaluation. This revealed deltoid and distal upper extremities myopathy mostly on the left with Neurosensorial Neuropathy of the left median and ulnar nerves, and bilateral radial nerves corresponding to Segmental Demyelination type and Axonal Degenration.

Case 2

A 29-year-old African American male presented with progressive worsening of bilateral hand and forearm weakness that was worse on the left. He also had mild difficulty walking due to weakness in his lower extremities. The patient reported an eight-year history of upper extremity weakness that had recently worsened during the winter months.. Patient denied any history of trauma to his neck. His family history was unclear as he was adopted.

On examination, he had a significant asymmetric atrophy of small muscle of hands and forearm with sparing of his brachioradialis and arm muscles. There were marked minipolymyoclonus observed in his hands He also had hyper-reflexia in his lower extremities, with sustained clonus of the left lower extremity.

Due to clinical findings suspicious for Hirayama disease, imaging studies were performed with T1- weighted Magnetic Resonance (MR) of the cervical region in neutral (Figure 2 A and B) and neck flexed position (Figure 2 C and D). T2-weighted MR (Figure 3A and B) was ordered for further evaluation.

Physical Exam

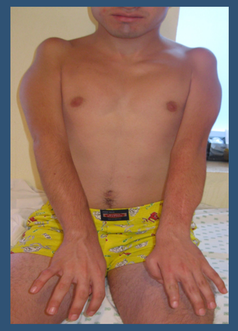


Figure 1A. Atrophy of left shoulder girdle.

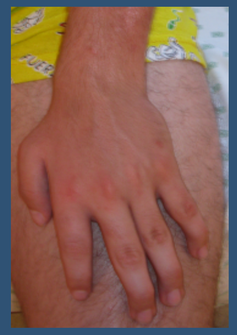


Figure 1B Asymmetric atrophy of third and fourth left dorsal interousseous muscle.

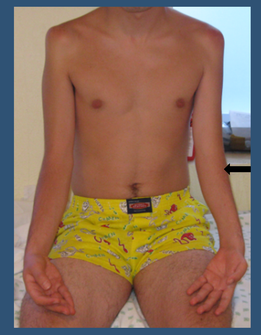
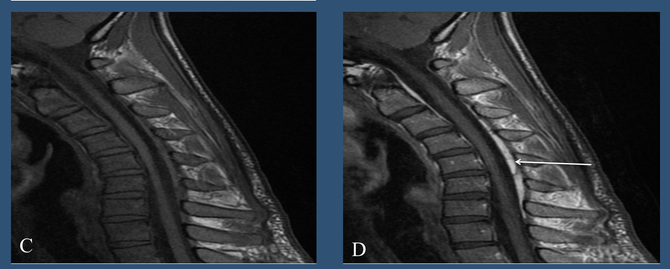


Figure 1C. Preservation of the brachioradialis mucle consistent with oblique atrophy. (arrow)

Imaging Studies

Figure 2. (A) Pre- and (B) Post-Contrast T1-weighted images of cervical spine in neutral position, (C) Pre- and (D) Post-Contrast T1 images of flexed cervical spine showing anterior displacement of posterior dura from C5 to C7 and enhanced posterior epidural space more prominent on flexion (arrows).



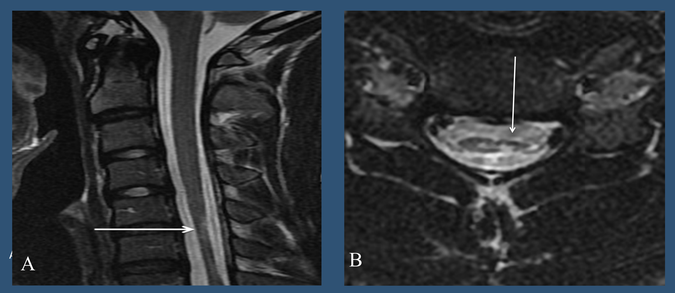


Figure 3. (A) Sagittal and (B) Axial T2-weighted images showing displacement of the posterior dura and focal hyperintensive signals in the left posterior region corresponding to the left dorsal column (arrows).

Discussion

Hirayama disease, also known as Juvenile Muscular Atrophy of Distal Upper Extremity, is a benign, uncommon disease predominantly seen in young men of Southeast Asian descent[1] It often presents with insidious onset of muscular atrophy of the hands and forearms, and generally spares the brachioradialis[1]. This is known as oblique amyotrophy.[1] Patients may also feel trembling and "cold paresis", or weakness of fingers when exposed to cold.

The primary predisposing factor is an imbalanced growth causing a tight dural sac.[2] When the neck flexes, the unusually tight dural sac cannot compensate for the increase in length of the posterior dural wall[2]. This causes the posterior wall to shift and compresses the cord against the vertebral bodies.[2] The chronic flexion of the neck likely causes micro-vascular ischemia to the anterior horn resulting in gliosis and atrophy of the cord. [1,2]

Hirayama disease is best characterized with MR of the cervical spine in neutral and flexed positions.[3] In the neutral position, MR demonstrates localized lower cervical cord atrophy, asymmetric cord flattening, and loss of attachment. [1,3] In flexed position, MR reveals anterior displacement of posterior dural sac, loss of dural attachment, and increased posterior epidural tissue containing serpiginous flow voids.[1,3] Focal gliosis and atrophic narrowing of the involved lower cervical cord can occur over time from chronic spinal cord impingement and persists on imaging in both flexion and neutral positions. [1,2] The serpiginous flow voids demonstrated on MRI corresponds with engorgement of the epidural venous plexus. While the epidural venous plexus engorgement is thought to be a passive and transient process in association with neck flexion, it may contribute to the anterior horn damage of the spinal cord. [2]

Application of a cervical collar for 3 to 4 years has been show to stop progression of the disease.[1]Another treatment alternatives include anterior cervical decompression, duraplasty, and reconstruction with tendon transfer. [1]

References

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