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Imaging in Hirayama Disease

Hirayama Hastalığında Görüntüleme

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Dear editor,

A 14-year-old boy presented with gradual muscle loss associated with his right hand and forearm weakness over 2 years, which later also involved his left hand. Additionally, he complained of fine tremors and inability to carry out daily activities. The patient did not report any relevant family or trauma history. Physical examination revealed significant muscle loss of the upper limbs except for the bilateral brachioradialis and left deltoid muscles. Moreover, a significant reduction was observed in the strength of the hypothenar muscles, finger extensors, and muscles innervated by the ulnar nerve. Electroneuromyography revealed normal sensory responses but decreased motor responses involving the ulnar, median, and radial nerves with denervation potentials in C7-T1 dermatomes. Sagittal T2-weighted magnetic resonance imaging (MRI) in the neutral portion of the neck showed mild volume loss of the spinal cord at the C6-C7 levels. When MRI was repeated in the flexion position of the neck, anterior displacement of the posterior dura with spinal cord compression was observed. Additionally, the widening of the posterior epidural space was observed with prominent flow voids (Figure 1).

Hirayama disease (HD), also known as non-progressive juvenile spinal muscular atrophy, commonly affects the upper limbs and typically spares the brachioradialis muscle. HD is characterized by asymmetrical weakness and muscular atrophy predominantly affecting the C7, C8, and T1 myotomes. It occurs in males aged 15-20 years (1,2,3). Almost all patients present with unilateral

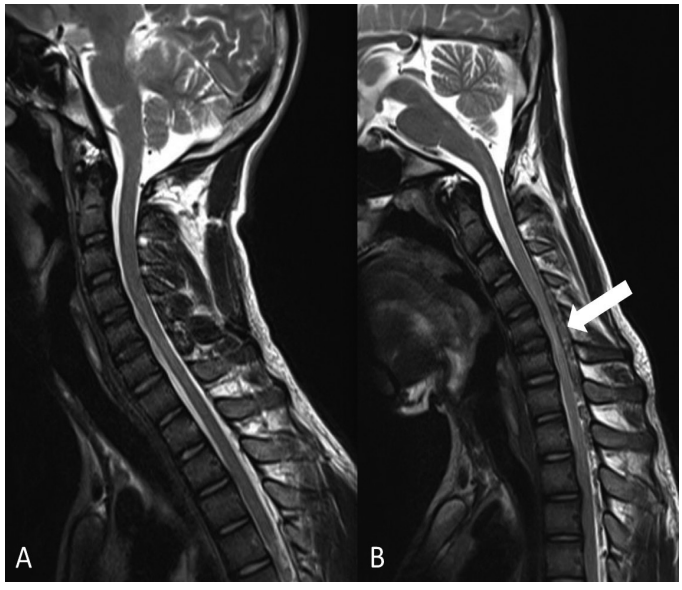

Figure 1. Sagittal T2-weighted magnetic resonance imaging in (A) neutral position of the neck reveals a mild volume loss of the spinal cord at the C6-C7 levels and (B) in flexion position reveals an anterior displacement of the posterior dura with spinal cord compression (white arrow) and widened posterior epidural space with prominent flow voids

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upper limb involvement, but some cases of bilateral upper limb weakness have also been reported as in our case. Disease mechanism and pathogenesis are unknown. Usually, the dura surrounding the spinal cord is larger for its contents and is attached at the foramen magnum, posterior longitudinal ligament at the C2-C3 vertebrae (1,2,3). In HD, an abnormal anterior displacement of the posterior dural wall results in cord compression. The imbalance between the growth of the vertebral column and its content results in abnormal dural sheath tightening (1,3). During neck flexion, spinal canal narrowing occurs, thus the spinal cord undergoes multiple episodes of trauma and ischemic insults resulting in myelopathy. MRI done in neck flexion shows anterior displacement of the posterior dural wall along with compression and decreased anteroposterior diameter of the cord with posterior epidural space enlargement, which appears as crescent-shaped high T1 and T2 signal intensities (2,4,5). HD differentials include amyotrophic lateral sclerosis, spinal muscular atrophy, brachial plexopathy, and syringomyelia. HD is differentiated from these conditions by its clinical, electrophysiological, and radiological characteristics (4). Early detection of HD prevents further disease progression. In the initial stages, the patient is advised to use the cervical collar to limit neck flexion and prevent further spinal cord trauma. Decompressive surgery is performed in cases of persistent neurological deficit (3). In our case, the patient was advised to use a cervical collar and was followed up for disease progression monitoring.

Ethics

Informed Consent: Informed consent was obtained from individual participant included in the study.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: N.K.G., N.K., D.E., Concept: N.K.G., N.K., D.E., Design: N.K.G., N.K., D.E., Data Collection or Processing: N.K.G., N.K., D.E., Analysis or Interpretation: N.K.G., N.K., D.E., Literature Search: N.K.G., N.K., D.E., Writing: N.K.G., N.K., D.E.

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