Clinical Image

**Flexion-Induced Cervical Cord Compression: Hirayama Disease**

**Tolu et al. Hirayama Disease**

Sena Tolu¹, Fikret Aysal², Tuğrul Örmeci³, İbrahim Ethem Kirez³, Nurbanu Hindioğlu¹

¹Department of Physical Medicine and Rehabilitation, Medipol University School of Medicine, Istanbul, Turkey
²Department of Neurology, Medipol University School of Medicine, Istanbul, Turkey
³Department of Radiology, Medipol University School of Medicine, Istanbul, Turkey

**Address for Correspondence:** Sena Tolu, Department of Physical Medicine and Rehabilitation, Medipol University School of Medicine, Istanbul, Turkey
Phone: +90 505 442 47 22 e-mail: dr.sena2005@gmail.com ORCID: 0000-0002-1111-3110

Received: 5 January 2019
Accepted: 20 February 2019

**DOI:** 10.4274/balkanmedj.galenos.2019.2019.1.16

Cite this article as:

A 15-year-old male presented with progressive weakness and amyotrophy of the right distal arm and hand muscles, which he had been present for the last three months. The patient had no comorbid diseases and history for cervical trauma. His family members had no neuromuscular disorders. A clinical examination showed weakness of the right interossei muscles [grade 3/5 on the Medical Research Council (MRC) scale], abduction of the right thumb, and extension of the right wrist and fingers II-V (MRC 4/5). On inspection, there was marked atrophy of the right first dorsal interosseous muscle and mild atrophy of other intrinsic hand, flexion and extension muscles of the wrist. There was no fasciculation, sensory deficit, and pain, but tremulous movement of his fingers was observed. Deep tendon reflexes were intact. A neurologic examination of the left upper and lower extremities was normal and no signs of pyramidal tract involvement were present. Babinski reflex and Hoffman’s sign were negative. Motor nerve conduction studies showed a reduced amplitude of compound muscle action potential of the right ulnar nerve but normal parameters of the left ulnar, and bilateral median nerves. No focal slowing or conduction block were found. Sensory nerve conduction studies were normal. An electromyographic (EMG) examination found active denervation in the right C8-T1 and C7 innervated muscles (abductor pollicis brevis, first dorsal interosseus, abductor digitii minimi, extensor digitorum communis, flexor carpi radialis, triceps muscles). A pure motor deficit affecting roots C7 to T1 was diagnosed. Blood analyses were normal. Cervical vertebra magnetic resonance imaging (MRI) performed in the neutral position was normal (Figure 1a-b). On the basis of clinical, EMG and MRI findings, we ruled out cervical cord pathologies, brachial plexopathy, multifocal motor neuropathy with conduction block, spinal muscular atrophy and amyotrophic lateral sclerosis. On the suspicion of Hirayama disease (HD), MRI acquired in full flexion of the neck revealed dilation of the posterior internal vertebral plexus due to forward displacement of posterior dura mater (Figure 1c-f). The widening of the posterior epidural space disappeared in the neutral position. He was diagnosed to have HD and treated conservatively. He was recommended to wear a cervical collar to minimize neck flexion.

HD is a rare and a self-limited condition characterized by asymmetric muscle weakness and amyotrophy of the distal upper limbs involving the C7-T1 myotomes. The pathologic findings implicate circulatory changes in the lower cervical cord which causes ischemic changes in the anterior horn. Congestion of the venous plexus, abnormal drainage in the vertebral venous plexus or epidural vascular malformation may be responsible (1,2). In this case, the diagnosis is guided mainly by electrophysiological and flexion MRI findings.

**REFERENCES**

Figure 1. Hirayama disease. Normal MRI finding on sagittal T2-weighted images in a neutral position (a) and in slight flexion (b). However, dolichoectatic vascular structures (arrows) are seen on the sagittal T1-weighted image (c), sagittal T2-weighted image (d), axial T1-weighted image (e) and axial T2-weighted image in increased flexion. Prominent posterior epidural space and compression of the spinal cord are present between C5-D1 levels (c-f).