Cervical Myelopathy due to a “Tight Dural Canal in Flexion” with a Posterior Epidural Cavity

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Abstract

A 41-year-old man noticed weakness and atrophy in his right hand and forearm resembling the non-progressive juvenile muscular atrophy of unilateral upper extremity (Hirayama’s disease). MRI showed an abnormal cavity in the posterior epidural space which appeared on neck flexion communicating with the subarachnoid space in addition to the flattening of the lower cervical spinal cord on neck flexion. When evaluating atypical cases of Hirayama’s disease, the pathomechanism demonstrated in the present case should be taken into consideration.

Key words: flexion myelopathy, Hirayama’s disease, muscle atrophy

Case Report

A 41-year-old man was admitted to our hospital complaining of weakness in his right hand. In March of 1992, he experienced several transient attacks of severe pain in the posterior neck, which were relieved by lying down. In July of the same year, the weakness in his right hand abruptly appeared after an attack of severe nuchal pain, resulting in decreased grip strength and clumsiness. Partial recovery and exacerbation were alternately repeated. His symptoms gradually worsened. He was evaluated by an orthopedic surgeon at a local hospital and was diagnosed as having cervical spondylodiscitis for which surgery was recommended. In October of 1997, he was transferred to our department for further evaluation and with a suspicion of Hirayama’s disease.

A neurological examination revealed 1) weakness and atrophy of the right wrist flexor and intrinsic hand muscles; 2) fasciculations in the right intrinsic hand muscles; and 3) decreased bilateral triceps reflexes. Cerebrospinal fluid analysis was normal. Nerve conduction velocities and SEP of both median and ulnar nerves were within normal limits. A needle EMG showed giant complexes at both musculi extensor carpi radialis. In MEP studies, amplitude was transiently elevated after neck flexion, which was followed by low amplitude, long duration, and polyphasic MEPs after seven minutes. When his neck was flexed, the patient had difficulty moving all of the fingers of his right hand, but his symptoms were reduced when his neck was restored to its neutral position.

MRI of the cervical spine revealed a mild bony spur and disk herniation, as well as lower cervical spinal cord atrophy, and there was high signal intensity in the cord at C5/6 level on T2WI (Fig. 1A). On neck flexion, the lower cervical spinal cord was found to shift anteriorly and an abnormal cavity became evident in the posterior epidural space (Fig. 1B). This abnormal epidural cavity showed high signal intensity on T2WI and low signal intensity on T1WI, suggesting that it was filled with fluid. The lower cervical spinal cord was pinched between the vertebral bodies and the posterior wall of the dural tube. Myelography showed stasis of contrast medium with clear boundaries at the back of the spinal cord (Fig. 2A), and leak-
Tight Dural Canal in Flexion

Figure 1. Cervical spinal MRI (T2WI). A) Sagittal view, neutral position. The lower cervical spinal cord was atrophic. A disc herniation was found at C5/6, and high signal intensity was shown at the C5/6 spinal cord level. B) Sagittal view, flexed position. The lower cervical spinal cord was shifted anteriorly and an abnormal cavity became apparent in the posterior epidural space. It was also pinched between the vertebral bodies and the posterior dura.

Discussion

Since the first reported description of Hirayama’s disease in 1959 (1), there have been several reported cases of a juvenile type of distal and segmental muscular atrophy of the upper extremities (4). Those cases are now thought to have belonged to the clinical entity known as Hirayama’s disease, whose criteria were listed above. In 1987, a neuropathological examination of the first autopsy case was performed, suggesting circulation insufficiency of the lower cervical spinal cord as the cause of the disease (5).

The patient described in this report had many symptoms that were characteristic of Hirayama’s disease, but also displayed some atypical features such as an older age of onset and severe nuchal pain, the latter of which was a rare finding. He also had an abnormal epidural cavity which, in all likelihood, contributed to his symptoms. Though disproportionate growth of the spinal column, spinal cord, and dural tube was thought to be the most direct cause of the symptoms of Hirayama’s disease, our patient had already reached his full height and as such, his symptoms were unlikely to have been due to a developmental disorder.

We previously introduced the term “flexion myelopathy and tight dural canal in flexion” (3, 4) to refer to all myelopathies caused by constant pressure on the cervical cord as a result of neck flexion. Our emphasis was on dynamic rather than static factors as the basis for the disease. It has been speculated that disproportionate development of the bony spinal canal and spinal cord causes “over-stretching” of the cervical cord in this condition (3, 4). However, in our case of an adult who was fully grown when the disease manifested itself, the cause of detachment of the dural tube from the spinal canal resulting in the development of an abnormal epidural space (due to communication with the subarachnoid space) is unknown. The shifting of this cavity forward could have caused spinal cord compression by the same mechanism as described above.

It has previously been reported that the expanded epidural space that develops in Hirayama’s disease results from dilation of the spinal venous plexus. The existence of an abnormal epidural cavity, such as described in the present patient, has not been previously reported in Hirayama’s disease but should be considered as a possible mechanism in patients with atypical disease.
Figure 2. A) Myelography. In a sagittal view on neck flexion, stasis of contrast medium with clear boundaries was demonstrable at the back of the spinal cord. B) CT myelography in neutral position. Contrast medium was recognized in both the subarachnoid space and the epidural cavity. The subarachnoid space around the spinal cord was normal. C) CT myelography on neck flexion. The subarachnoid space around the spinal cord was narrowed. The spinal cord was shifted anteriorly and flattened. An expansion of the epidural cavity was recognized on neck flexion.

References


