Dissociated Motor Loss Syndrome with Cavities in the Anterior Horns

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A 74-year-old man developed proximal muscular weakness and wasting of the left upper extremity without sensory disturbance or myelopathic symptoms. The muscle atrophy had not progressed for a few years. Radiological examination of the spine showed cervical disc herniation. These findings and electrophysiological studies excluded motor neuron diseases, permitting the diagnosis of dissociated motor loss syndrome. Interestingly, delayed computerized tomographic myelography disclosed cavities in the anterior horns of the spinal cord, which coincided with the clinical symptoms. Previous radiological and pathological examinations showed formation of such cavities within the spinal cord resulting from chronic compression, which was followed by ischemic change. In this context, the present case supports ischemia as a cause of dissociated motor loss syndrome.

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Introduction

Dissociated motor loss syndrome was first described by Keegan (1). This syndrome, which is almost the same clinical entity as cervical spondylotic amyotrophy described by Sobue et al (2) and Yanagi et al (3), is a unique disorder of the cervical spine, characterized by marked muscle wasting of the upper extremity without sensory disturbance or myelopathy.

These neurological manifestations may be mistaken for motor neuron diseases (MND). However, the clinical course, clinical examinations such as deep tendon reflexes, and electrophysiological studies may be useful in differentiating this syndrome from MND.

Compression of the anterior root of the spinal cord by spondylotic spine or herniated disc is thought to cause this syndrome (1). Sobue et al (2) and Yanagi et al (3), however, emphasized the importance of blood flow disturbance caused by chronic compression. The present case is believed to support their hypothesis.

Case Report

A 74-year-old man was admitted to the hospital with muscular wasting and weakness around the left shoulder. The patient had a long history of chronic rheumatoid arthritis (RA). Five years earlier, he began to notice muscular weakness around the left shoulder. The muscular weakness was followed by muscular wasting and gradually worsened. However, this muscle wasting had not progressed recently. No sensory disturbance was present. The muscular weakness and wasting revealed marked laterality, dominating on the left side. He had been a farmer and had no history of traumas and traffic accidents.

Before admission, he had been treated with prednisolone for RA. Muscular weakness improved transiently during steroid hormone therapy, but worsened with difficulty in lifting his left arm when the dose was tapered.

On physical examination, the cranial nerves were intact. No atrophy or fasciculation was seen in his tongue. Muscular wasting was seen around the shoulder. Manual muscle testing revealed localized muscle weakness and was graded as 4/5 in the left deltoid muscle, 4+/5 in right and 3+/5 in left biceps brachii muscles, and 3+/5 in left triceps muscle, although no muscle weakness was seen in the legs. Fasciculation was detected in the left triceps muscle, but was not seen in the lower extremities. Deep tendon reflexes were absent in the biceps and triceps, but were normal in the knees and ankles. Plantar responses were flexor. No sensory disturbance or ataxia was seen. Urination was normal.
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Figure 1. Magnetic resonance images of the neck. Cervical disc herniation was observed at the level of C3-C6. Left) sagittal T1 weighted image (spin-echo sequence with a repetition time of 300 milliseconds and an echo time of 25 milliseconds, 0.5-T Magnetom). Right) sagittal T-2 weighted image, (spin-echo sequence with a repetition time of 2000 milliseconds and an echo time of 40 milliseconds, 0.5-T Magnetom).

Routine laboratory studies were normal except for mild elevation of creatine kinase. Cerebrospinal fluid examination was normal. Needle electromyogram study revealed positive sharp waves in the left deltoid and biceps muscles, but no abnormality in the muscles of the paraspinal area or lower extremities.

Myelography and magnetic resonance imaging (MRI) of the neck (Fig. 1) showed cervical disc herniation, which compressed the spinal cord at C3/4, C4/5, C5/6 and C6/7. Delayed computerized tomographic myelography (Fig. 2) and MRI of the cervical cord disclosed a flattened cord at C3/4 and C4/5, and cavities in the anterior horns of the spinal cord at C-4, C-5 and C-6.

Steroid therapy was no longer effective. He was able to take care of himself, and refused any operations for the cervical disc herniation. After discharge, the muscle weakness and wasting showed no significant progression and motor strength in the legs maintained normal for next one year.

**Discussion**

The present case showed localized muscular wasting, which was non-progressive for several years, without sensory disturbance. Electromyogram revealed denervation activities in the atrophic muscles. Radiological examinations disclosed cervical herniation which compressed the spinal cord. MND was ruled out by the long clinical course, symptoms and electrophysiological study, permitting the diagnosis of dissociated motor loss syndrome.

Keegan first described this syndrome (1). In his report, he noted flattening of a cervical nerve root due to large spondylosis. He suggested that the dissociated motor loss in the upper extremities was due to discrete compression of the motor roots intradurally by posterolateral spondylosis. He also showed normal anterior columns and horns, excluding cystic degeneration as a cause of this unique syndrome.

However, not all the clinical symptoms coincide with the level of spondylosis or disc herniation, supporting ischemia of the anterior horn as a cause of this syndrome rather than compression of the motor root alone.

Iwasaki et al (4) described a high density area that resembled “fried eggs” in the gray matter of the spinal cord near the abnormal cervical disc in delayed CT myelography. It was of interest that delayed CT myelography of the present case also
disclosed a “fried eggs” appearance in the anterior horn of the spinal cord, which coincided with the clinical symptoms. Takahashi et al (5) and Matsuda et al (6) also showed the increased magnetic resonance signal intensity, some areas of which showed microcavities, within the spinal cord due to chronic cervical cord compression.

Pathological change of the spinal cord caused by chronic compression has been described previously (7–9). Hashizume et al (7) disclosed that chronic compression by a cervical lesion affected the gray matter more severely than the white matter and formed cavities within the spinal cord. In the present case, steroid therapy was effective transiently, indicating the presence of edema within the spinal cord resulting from ischemic change.

Iwasaki et al (4) and Mair and Druckman (10) suggested that chronic compression of the spinal artery by a cervical lesion leads to ischemia, resulting in the formation of the intra-spinal cavity. On the other hand, Taylor and Byrnes (11) showed that anterior horn cell damage below the level of cord compression occurred from hypoxia secondary to interference with venous drainage by the compressive lesion (12). At present, no arterial or venous dilatation is detectable in MRI of the present case, but the spinal cord above the level of cavities at C5 was flattened although no deformity of spinal cord was seen at C5. In this context, the present case supports Sobue and Yanagi’s ischemic theory as a cause of dissociated motor loss syndrome (2, 3).

References