Spinal Segmental Myoclonus during Postural Maintenance in a Patient with Cervical Spondylosis: A Case Report

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Abstract

Spinal segmental myoclonus is defined as a rare involuntary movement characterized by myoclonic jerks of spinal origin. We describe the case of a 62-year-old woman who developed spinal segmental myoclonus 4 months after undergoing cervical laminoplasty for ossification of the posterior longitudinal ligament. Myoclonic jerks were observed in the upper trapezius innervated by C3-4, which corresponded to the level of myelomalacia. These jerks were elicited and aggravated in the sitting and standing positions but were completely suppressed in the supine position. The myoclonus was refractory to medication but improved with the use of a soft neck brace.

Key words: spinal segmental myoclonus, reticulospinal tract, antigravity muscles, spinal inhibitory interneurons


Introduction

Myoclonus is defined as a sudden, shock-like involuntary movement due to muscle contraction or inhibition. It may be caused by lesions occurring at various levels of the central nervous system such as the cortex, subcortex, and spinal cord. Spinal segmental myoclonus (SSM) consists of myoclonic jerks of the muscles corresponding to the innervating spinal segments. Several causes of SSM have been described: tumors, infections, demyelinating diseases, vascular lesions, and spondylosis (1). SSM responds to treatment with agents such as clonazepam, trihexiphenidyl, carbamazepine, baclofen (1), and botulinum toxin type A (2). However, the alleviation of SSM by the application of neck brace therapy has not been reported to date. We report the case of a patient who developed SSM after undergoing laminoplasty for cervical spondylosis and was successfully treated by the use of a soft neck brace.

Case Report

A 62-year-old Japanese woman underwent cervical laminoplasty between C3 and T1 for cervical spondylosis caused by ossification of the posterior longitudinal ligament. Four months after surgery, the patient developed involuntary myoclonic elevation of the left shoulder that presented only in the sitting and standing positions. Neurological examination indicated muscle weakness in the left upper extremity (grade 4 on the Medical Research Council scale). Deep tendon reflexes were slightly brisk in all extremities and pathological reflexes were negative. The patient experienced paresthesiae in the entire palmar region and left plantar region. Her coordination, cognitive functions, and other neurological findings were normal. The involuntary, sudden, and comparatively rhythmical movements were observed only in the left upper trapezius, with a frequency of about 70 jerks per minute. These abnormal contractions were not induced by any other stimulus such as large noises and tendon taps, but were exacerbated by emotional stress and specific body positions requiring postural maintenance. Motor and sensory

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nerve conduction studies of the median, ulnar and posterior tibial nerves were normal, and the long-loop response was negative. Somatosensory evoked potentials (SEP) recorded after the median and posterior tibial nerve stimulation did not show large cortical SEP. The jerk-locked back averaging technique displayed no cortical electrical activity over the scalp prior to the myoclonic jerks of the upper trapezius. No abnormal lesions were detected on brain magnetic resonance imaging (MRI) and single photon emission computer tomography. Cervical MRI demonstrated intramedullary high intensities on T2-weighted images at the C3-4 level predominantly in the left posterior horn. This finding was consistent with myelomalacia (Fig. 1A, B). Surface electromyograms were recorded from the sternocleidomastoid, posterior neck muscles, upper and lower trapezius muscles, deltoid, biceps and triceps. On electromyographic examination, there were no abnormal discharges in the supine position (Fig. 2A). However, 0.9-1.3 Hz rhythmic groups of discharges lasting approximately 170-500 ms were recorded only in the left upper trapezius, immediately after assuming a sitting or standing position (Fig. 2B, C). The patient had been treated with several medications such as clonazepam, valproate, carbamazepine, baclofen, trihexyphenidyl, and intramuscular injections of botulinum toxin type A into the upper back for 15 years since the onset of SSM. However, satisfactory results were not obtained. The clinical and physiological characteristics of SSM remained almost unchanged over this period of time. Myoclonus was not alleviated by applying tactile stimuli such as placing a scarf around the patient’s neck. However, an immediate marked reduction in the frequency and amplitude of the groups of discharges was obtained by providing adequate neck support by either placing hands under the mandible or using a soft neck brace (Fig. 2D). The use of a soft neck brace significantly reduced the extent and frequency of SSM.

**Discussion**

The myoclonus observed in the left shoulder was thought to be of spinal origin because it was localized in the upper trapezius innervated by C3-4 segments that were shown to have myelomalacia on cervical MRI. There were no electrophysiological findings indicating a cortical origin of the myoclonic jerks, including a large SEP, a cortical long-loop reflex, and a sharp wave preceding the myoclonus on applying the jerk-locked averaging technique. The pathophysiology of SSM remains unclear; however, two pathological mechanisms of SSM have been previously proposed. First, Shivapour and Teasdall suggested that spinal myoclonus may be caused by increased excitability of anterior horn cells at the time of sublethal injury (3). Secondly, Davis et al demonstrated that abnormal alpha motor neuron excitability resulting from a severe reduction in the activity of inhibitory intermediate neurons in the posterior horn could induce myoclonus and rigidity (4). Moreover, intramedullary lesions represented by syringomyelia often lead to several
types of spontaneous activities such as continuous motor unit potentials, synchronous motor unit potentials, and spinal myoclonus (5, 6). In the present patient, long-term clinical stability of SSM and cervical MRI findings suggest that the disinhibition of alpha motor neurons resulting from the activities of spinal interneurons may be associated with the development of SSM.

The reticulospinal system controls and coordinates postural movements of the entire body and contributes to the regulation of axial and postural muscle tone (8). In addition, reticulospinal terminals are distributed mainly among intermediate zone interneurons or medial motor neurons that innervate the axial and proximal limb muscles (8). We speculate that the spontaneous activity of isolated alpha motor neurons, along with an increase in trapezius muscle tone mediated by the reticulospinal tract during postural maintenance, may lead to the development of myoclonic jerks in the sitting and standing positions. Based on this speculation, it was hypothesized that supporting the neck with a soft brace, which would also support antigravity muscles, may alleviate facilitation of the reticulospinal system in addition to modulating proprioceptive afferent inputs (7). These mechanisms may provide relief from myoclonic jerks in some SSM patients.

The authors state that they have no Conflict of Interest (COI).

References