Painful Spasms and Rigidity of the Lower Limb Following Transverse Myelitis Associated With Sjögren’s Syndrome

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Abnormal muscle tone, such as spasms, rigidity, and stiffness, following acute transverse myelitis (ATM) was such a rare manifestation that hardly reported until now. We experienced a 50-year-old patient with ATM associated with Sjögren’s syndrome. Furthermore, the patients complained painful spasms and rigidity of left lower limb which begun after episode of ATM. Journal of Movement Disorders 1(1):51-54, 2008

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Acute transverse myelitis (ATM) is an inflammatory process that involves a restricted area of the spinal cord and it can involve the entire thickness of the spinal cord. It is a rare event with an estimated incidence of 1.34 per million in the general population. However, ATM may occur in about 1% of patients suffering with Sjögren’s syndrome. We experienced an infrequent case of ATM associated with Sjögren’s syndrome. Furthermore, the patient had painful spasms and rigidity of the lower limb following ATM. The spasms, which were spontaneous, stimulus-sensitive and induced by voluntary action, involved an abnormal pattern of muscle activity that consisted of repetitive grouped discharges of the motor units. There are relatively few reports of spinal cord disease causing these movement disorders as manifestation of the side effects of paraparesis. Therefore, we report here on an infrequent case with painful spasm and rigidity of lower limb following ATM accompanied with Sjögren’s syndrome.

CASE REPORT

A 50-year-old woman was admitted with painful spasms and rigidity of the left low limb that was induced by voluntary action. She had been diagnosed with ATM associated with Sjögren syndrome about 60 days previously. At that time, she developed ATM with paresthesia below the waist, an improper gait due to paraparesis and jerkiness of the legs on standing further, and urinary difficulty due to sphincter disturbance. These symptoms had progressed over a period of about one month. Thereafter, the sphincter disturbance had resolved and the paresthesia and paraparesis had slowly improved, but the patient did not fully recover. Subsequently, she was able to walk unaided with mild gait disturbance. The Shirmer test, which was performed because of her complaints of dry eye and dry mouth, demonstrated positive findings and the results of laboratory test showed positive findings for anti-ds-antibody, anti SS-A (Ro) and anti SS-B (La), yet the folate, vitamin B12, complement components C3 and C4, antcardiolipin antibody, lupus anticoagulant, rheumatoid factor and C-reactive protein were all normal. The salivary grand scan demonstrated decreased salivary gland function (Fig. 1-A,B). Thus, she was also diagnosed as having Sjögren’s syndrome on the basis of the above-mentioned results and her clinical symptoms. The
Salivary gland scans and thoracic spine magnetic resonance imaging of the patient. (A) On the baseline study, the salivary activity was diminished in the parotid gland and possibly the submandibular gland. (B) After stimulation with lemon juice stimulation, the salivary activity became intense. (C) On sagittal T2-weighted magnetic resonance imaging, diffuse signal intensities were seen in the spinal cord from the C7 level to the T10 level, and mainly at T5-T8. (D) On the axial T2-weighted magnetic resonance imaging, high signal intensities were seen in the dorsolateral part of the involved central gray matter of the 7th thoracic cord level.

magnetic resonance imaging (MRI) of the brain was unremarkable, but the sagittal T2-weighted MRI of the thoracic spine showed high signal intensities in the upper to middle thoracic cord (mainly at T5-T8 levels) (Fig. 1-C,D). This signal change involved the central and laterodorsal regions of the thoracic cord on the axial images. Her neurological condition then remained stable until about 25 days later.

On this admission, her painful spasms and rigidity of the left leg occurred whenever she move her left leg and foot. Neurological examination showed improved neurological symptoms, including paraparesis, paresthesia below the waist and sphincter disturbance, as compared with her previous condition, except for the painful spasms and muscle rigidity, which occurred on voluntary or passive movements of her left leg and foot. There were no other neurological deficits. On the electromyography test that was performed to evaluate the painful spasms of her left leg, continuous motor unit activity was recorded at rest in the tibialis anterior and gastrocnemius muscles of the left side (Fig. 2). There was no resting activity in the trunk muscles. Voluntary activity and passive movement of the left leg and foot precipitated the spasms and rigidity in the leg muscles. These spasms lasted several seconds and they were
exacerbated on resting activity. According to the above mentioned results and clinical symptoms, we diagnosed that the muscle spasms and rigidity was not a recurrence of ATM but a complication of ATM. Her painful spasms and rigidity mildly resolved by oral baclofen and clonazepam, but there was no remarkable improvement.

DISCUSSION

There are relatively few reports concerning spinal cord disease that causes spasms and rigidity, continuous motor unit activity, and reflex-induced and action-induced spasm of the lower limbs. In most reports, such pathology as intrinsic tumor, syringomyelia, vascular insufficiency or paraneoplastic myelitis has preferentially involved the gray matter. However, our present patient had ATM of middle thoracic level as the cause of her painful spasms and rigidity, and also had Sjögren syndrome in which ischemia due to vasculitis could be important for the genesis of myelitis. But the pathogenesis of transverse myelitis in rheumatic disease, such as Sjögren syndrome, is still unclear. According to previous report, only vasculitis and the immunological reaction of antineuronal antibody have been suggested as possible causes.

The mechanism of these manifestations was uncertain in the previously reported cases reference. Based on the clinical similarity among the prior patients who had known focal pathology that preferentially involved the gray matter of the spinal cord, disinhibition may have been caused by chronic focal spinal interneuronitis. Another report explained the mechanism for occurrence of abnormal muscle tone such as spasms, stiffness and rigidity following spinal cord injury. The normal flow of nerve messages below the level of injury is interrupted, and those messages may not reach the reflex center of the brain; the spinal cord then attempts to moderate the body’s response, but because the spinal cord is not as efficient as the brain, the signals that are sent back to the site of the sensation are often over-exaggerated. An overactive muscle response is referred to clinician as spastic hypertonia, that is uncontrollable jerking movement, stiffening or straightening out of muscles, shock-like contractions of a muscle or group of muscles, and abnormal tone in the muscles. We thought that overactive muscle responses of our patient with myelitis were compatible with the above-mentioned prior reports, because our patient also suffered painful spasms and rigidity following middle spinal cord lesions, which can disrupt normal flow of nerve messages.

Treatment of overactive muscle tone may include injection of muscle relaxant agents and also medications such as...
baclofen, diazepam or clonazepam. Some patients suffering with severe spasms utilize a baclofen pump, which is a small, surgically implanted reservoir that applies drug directly to the area of spinal cord dysfunction. Sometimes, selective dorsal rhizotomy may be considered if the spasms interfere with sitting, bathing or general care taking. In case of our present patient, we utilized baclofen and clonazepam as an oral agent for treating the painful spasms and rigidity. The patient’s symptoms slightly resolved after treatment, but the symptoms did not completely resolve.

In conclusion, we report here on a rare patient with painful spasms and rigidity in her left lower limb as complications induced by ATM at the middle thoracic cord level. Furthermore, the present patient had ATM associated with Sjögren syndrome. We also assert that the present case provides further evidence that this kind of movement disorder may be caused by spinal cord pathology.

REFERENCES


