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Focal Hand Dystonia as a Sign of Demyelinating Attack in Multiple Sclerosis: Report of Three Cases

Multipl Sklerozda Demiyelinizan Atak Bulgusu Olarak Fokal El Distonisi: Üç Olgu Sunumu

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Summary

Although it is known that dystonia is a basal ganglia disease, dystonic symptoms have been observed in association with lesions of various sites located in sensory and motor pathways. We report three cases of paroxysmal focal hand dystonia, which may be due to the damage of the somatosensorial pathways in the cervical spinal cord. We suggest that the dystonia in our patients may be related to these active demyelinating cervical plaques. Two female and one male patients with definite relapsing remitting Multiple sclerosis (MS) between the ages of 22 to 45 were admitted with serious disability while using their right hands. In all three cases abnormal posture in the right hand and involuntary sustained contractions together with minor choreiform movements of the fingers were observed. Cervical MRI showed contrast-enhancing demyelinating lesions at the level of C2-3 in all patients. One of the patient's cranial MRI revealed also two new contrast-enhancing plaques on the neighbourhood of right posterior lateral ventricle and parietal cortex. No new or enhancing lesion was detected in the basal ganglia; indicating that the cervical spinal cord lesions were responsible for hand dystonia. In one of the patients, the right median somatosensory evoked potential response was absent in accordance with the clinical symptom. All three patients were treated with 1 gr. intravenous methylprednisolone per day for 5-10 days. Approximately one month later, clinical symptoms have been completely disappeared and control cervical MRI revealed resolution of the active lesions in all. (Turkish Journal of Neurology 2014; 20:141-143)

Key Words: Multiple sclerosis, focal hand dystonia, cervical demyelinating lesion

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Özet

Distoninin bazal ganglion hastalığı olduğu bilinmesine rağmen, distonik semptomlar sensoriyal ve motor yolların farklı bölgelerindeki lezyonlara bağlı olarak da gelişebilir. Bu makalede servikal spinal kord da somatosensoriyal yolların hasarına bağlı olduğu düşünülen paroksismal fokal el distonisi ile başvuran üç multipl skleroz (MS) hastasını sunmaya değer bulduk. Hastalarımızdaki distonin aktif demiyelinizan servikal plaklar ile ilişkili olduğunu düşündük. Kesin relapsing remitting MS tanılı 22-45 yaş arası iki kadın ve bir erkek hasta sağ ellerini kullanmada güçlük şikayeti ile başvurdu. Her üç olgunun da sağ elinde ısrarlı kontraksiyonlar ile anormal distonik postür ve parmaklarında minör koreiform hareketler vardı. Olguların servikal MRG tetkikinde C2-3 düzeyinde kontrast tutulumu olan demyelinizan lezyonlar gözlendi. Bir hastanın kraniyal MR tetkikinde sağ lateral ventrikül arka bacağı komşuluğunda ve sağ parietal kortekste 2 yeni kontrast tutan plak izlendi. Bazal gangliada, yeni ya da kontrast tutulumu olan plak saptanmaması nedeniyle el distonisinden servikal spinal korddaki lezyonun sorumlu olabileceği düşünüldü. Bir hastada klinik semptom ile ilişkili olarak sağ median somatosensoriyal uyartılmış potansiyel yanıtı elde edilemedi. Tüm hastalar 5-10 gün boyunca 1 gr intravenöz metilprednizolon ile tedavi edildi. Yaklaşık 1 ay içinde şikayetleri tamamen düzelen hastaların kontrol MRG tetkikinde önceki aktif lezyonların tamamen gerilediği izlendi. (Türk Nöroloji Dergisi 2014; 20:141-143)

Anahtar Kelimeler: Multipl skleroz, fokal el distonisi, servikal demiyelinizan plak

Çıkar çatışması: Yazarlar bu makale ile ilgili olarak herhangi bir çıkar çatışması bildirmemişlerdir.

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Introduction

Movement disorders are rarely seen in multiple sclerosis (MS). The question of whether they should be regarded as symptoms of attack is controversial and it depends on certain conditions. In such patients, the most commonly seen movement disorder is tremor (25-58%) (1,2,3), followed by paroxysmal dystonia, ballism and chorea, paroxysmal kinesigenic dyskinesia, parkinsonism, myoclonus, hemifacial spasms and continuous facial myokymia, Tourette syndrome and complex hyperkinetic movement disorders (4). Dystonia types associated with MS are cervical dystonia, dystonic writer's cramp (5), hand dystonia, blepharospasm, oromandibular dystonia (6), generalized dystonia or hemidystonia (7). The lesion producing the clinical picture can be at the levels of cervical spinal cord, brainstem, cerebellum, cerebellar peduncles, thalamus, subthalamic nuclei, internal capsule or basal ganglia (8,9,10). In this report, we present 3 MS cases with demyelination plaques on their cervical cords, who presented with isolated focal hand dystonia as the only symptom of a new demyelination attack.

Case Presentations

Case 1

Twenty two years old female patient came to our clinic in January 2013 with numbness and pain on the right hand and toe, left hand and left side of the torso. Cranial and cervical magnetic resonance imaging (MRI) showed typical demyelination plaques while her cerebrospinal fluid (CSF) showed oligoclonal band pattern 2. Multiple sclerosis diagnosis was made and the patient's complaints were resolved after 1000 mg/day intravenous methylprednisolone (IVMP) for 3 days. She visited again after one month due to right arm numbness and involuntary jerks in her hand. In her neurological exam, her right hand had dystonia with depression on 4th and 5th digits and choreiform motions (Figure 1). Superficial sensory examination showed hypoaesthesia on the upper right extremity. Vibration and position senses were normal. Deep tendon reflexes (DTR) were normoactive. Cervical MRI showed contrast holding lesions posteriolaterally located at the level of C2-C3. Cranial MRI showed demyelinated plaques without contrast during the chronic process. Right median SEP response was not acquired. The patient's complaints were completely resolved after 6 days of 1000 mg/day IVMP.

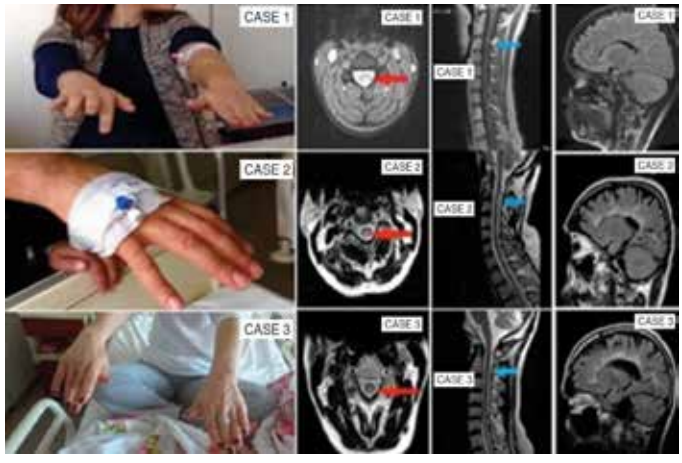

Figure 1. Dystonic hand postures of the cases and their MRI images highlighting the demyelinating plaques

Case 2

Thirty-seven-year-old male patient was being followed with MS diagnosis for 8 years. His complaints started in 2002 as loss of balance which disappeared in 2 days without treatment. He was diagnosed with MS due to hand numbness. He was given corticosteroid treatment due to the attacks involving his optical, cerebellar, pyramidal, sensory and bladder functions.

When he was using regular immunomodulatory treatment, he came to our clinic in March 2013 for involuntary convulsions and movements on his right hand fingers. His neurological exam showed dystonic posture and choreiform motion in the form of persistent abduction of the 5th digit of right hand. His surface sensory exam was normal while his position sense was reduced in lower right. His vibration sense was slightly reduced for four directions. Deep tendon reflex was strong on the right side and normoactive on the left. Cervical MRI showed demyelination plaques at the level of C2-C3 with peripheral contrast. Demyelinated plaques were present in many chronic processes without contrast as shown by cranial MRI. After 9 days of 1000 mg/day IVMP, his complaints were fully resolved.

Case 3

Fourty-five-year-old female patient was being followed with MS diagnosis fort he past fourteen years. Her symptoms first started as numbness of her left arm in September 1998. The patient who had sensory complaints which then improved without treatment was later diagnosed with MS in 1999. She was given corticosteroid treatment for the attacks involving cerebellar, optic, sensory and pyramidal functions.

She came to the clinic in March 2013 for the involuntary convulsions and movements for the past 4 days in her right hand as she was writing. Neurological exam showed dystonic posture of the 2nd right digit (Figure 1). Surface sensory study showed hypostesia on the right side. Position sense was normal. Vibration vibration sense was slightly reduced for four directions. Babinski and Hoffman signs and Achilles clonus were present. Bilateral cerebellar tests were moderately impaired. Deep tendon reflex was hyperactive for 4 sides. Cranial MRI showed chronic plaques in addition to 2 new contrast-holding demyelinated plaques on adjacency of the dorsal leg of lateral ventricle and parietal cotex. After 7 days of 1000 mg/day IVMP, his complaints were fully resolved.

Discussion

Dystonia is a neurological condition causing repetitive, twisting (contorting) motions or abnormal postures due to muscle flexion in the involved regions (11). Dystonia is known as a basal ganglia disease (12). Hand dystonia as an attack symptom in MS is rarely seen. The lesion causing the dystonia in MS and the relationship between these two conditions are not very well known. However, it is possible to establish a localization-attack relationship in patients showing cervical plaques concurrent with mono/hemidystonia; those who does not have basal ganglia lesions and who improve with IVMP. We also observed active demyelination plaques in the cervical MRIs of these 3 cases who came to our clinic with hand dystonia. All three of these patients responded to 5-10 day IVMP

treatment for the attacks, and the cervical plaque activity in the MRI was seen to subdue.

Uncini et al. Showed posterolateral cervical lesions using MRI in 2 MS patients who developed acute hand dystonia and athetoid movements (13). The mechanism causing the movement disorder was argued to be descending pathways controlling the reciprocal innervation of motor neurons and the involvement of large diameter afferents. Yücesan et al. discovered posterolateral and cervical C2-3 and C4 right posterolateral demyelination lesions in an MS patient with left hemidystonia in the absence of any basal ganglia lesions (7). It was hypothesized that the symptoms were due to cervical lesions when 1 gr pulse steroid administration caused the improvement in the cervical lesions as well as the hemidystonia in this patient. Schmidt et al. also reported dystonia and kinesigenic movements in 4 neuromyelitis optica cases who presented with cervical or cervicodorsal myelitis (14). In an MS case whose first ymptom was paroxysmal focal hand dystonia, Yonn et al. reported that the lesion responsible for the clinical picture was located in the dorsal leg of contralateral capsula interna (15).

In the literature, mostly in the form of case studies, movement disorders due to cervical cord lesions were also reported in syringomyelia (16), traumatic cervical spinal cord damage (12, 17), cervical cord tumor (18) in addition to demyelinating diseases. The demyelination plaques on the cervical spinal cord in the three MS cases presented here suggested that the dystonias may likely be due to the damage of the somatosensory pathways of the spinal cord. The lack of median SEP response in one of our cases can be an objective evidence for this hypothesis. Focal dystonia can be seen due to cervical cord lesions, and the fact that such movement disorders may develop due to acute demyelinating attacks in MS patients should be kept in mind.

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