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Spastic paraplegia as the only manifestation in neuropsychiatric lupus: a case report

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Abstract. – Demyelination syndrome is one manifestation of neuropsychiatric lupus erythematosus (NPLE) and is rare in systemic lupus erythematosus (SLE). When SLE presents only neuropsychiatric symptoms without damage to other systems, its diagnosis becomes difficult. Herein, we report a 29-year-old male who suffered from lower limb stiffness with recessive onset and progressive aggravation in one year. He had paraplegia, spastic hypertonia, abnormal gait, and bilateral positive Babinski signs. His symptoms indicated spastic paraplegia. Brain MRI showed multiple small demyelinating lesions in the lateral ventricle, brainstem, and cerebellum. Anti-ds DNA, anti-Sm and anti-RNP antibodies were positive. He was diagnosed with NPLE and had a good treatment response to steroids. To the best of our knowledge, this is the first case of spastic paraplegia as the only manifestation in SLE.

Key Words:
Systemic lupus erythematosus, Neuropsychiatric lupus erythematosus, Demyelination syndrome, Spastic paraplegia.

Introduction

Neuropsychiatric lupus erythematosus (NPLE) refers to nervous system impairment secondary to systemic lupus erythematosus (SLE), which manifests as a variety of psychiatric and neurological symptoms[1]. The American College of Rheumatology (ACR) has defined 19 types of NPLE, one of which is demyelination syndrome[2]. Patients with demyelination syndrome can present neuromyelitis optica (NMO), neuromyelitis optica spectrum disorders (NMOSD), demyelinating syndrome prominently affecting the brainstem, demyelinating syndrome prominently affecting the brain, and clinically isolated syndrome[3,4]. In addition to neuropsychiatric symptoms, patients with NPLE often have other systemic symptoms, such as skin damage, osteoarthritis, and blood system injuries[5]. However, when SLE presents only neuropsychiatric symptoms without damage to other systems, timely diagnosis could be difficult. If the patient has only atypical demyelination, diagnosis would become more difficult.

We hereby report a patient with SLE manifested as spastic paraplegia, a kind of upper motor neuron syndrome (UMNS), to help further understand the rare features of NPLE

Case Report

A 29-year-old male was admitted to our department with 1 year of experiencing stiffness in both legs. One year ago, he began to feel this stiffness in his legs without any inducement. His right leg was worse than his left leg. This stiffness would become apparent after a long period of standing or sitting or when waking up in the morning, and the stiffness was relieved after walking. His condition gradually got worse. Sometimes he had difficulty initiating walking. One month before, the patient gradually developed a stiff gait and was unable to stand. He could not move his legs when climbing stairs. His right leg involuntarily trembled at rest. He had no fasciculation, abnormal sensation, or bowel or urine dysfunction. His past history was normal. His family members did not have similar symptoms. His vital signs, including temperature, pulse, respiratory rate, and blood pressure, were in the normal range. There were no rashes on his skin. His cranial nerves were normal. The muscle strength of the upper limbs was normal, and

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muscle strength of the lower limbs was decreased by 4/5 degrees. His muscle tone was spasmodically increased in both arms and legs. Tendon reflexes were brisk on both sides. Bilateral ankle clonus was present. Coordinate movements were normal. Sensation was normal. Bilateral Babinski signs were positive. Routine blood tests, liver and kidney function, electrolytes and blood glucose were normal. Blood homocysteine was slightly increased to 15.9 μmol/L. Antiphospholipid antibodies were negative. The antinuclear antibody spectrum showed that the anti-ds DNA antibody, anti-Sm antibody and anti-RNP antibody were positive. The lumbar puncture cerebrospinal fluid (CSF) cell count was 11×10^6/L, and the protein concentration was 401 mg/L. CSF smear and acid-fast staining were negative. CSF culture, oligoclonal bands, AQP4 antibody and autoimmune encephalitis-related antibodies were negative. Chest CT showed no abnormalities. Peripheral nerve and spinal cord conduction velocities were normal. Brain and cervical T2 MRI sequences indicated hyperintense signals in the lateral ventricle, brainstem, and cerebellum, and cervical 3-4, 4-5, 5-6 intervertebral discs showed slight protrusion (Figure 1). Based on his medical history, positive antibodies related to SLE and abnormal brain images, the patient was diagnosed with NPLE and was given oral prednisone (50 mg per day). Two weeks later, his stiffness improved. His increased muscular tone returned to normal, and his walking was better than before. Currently, the patient lives like a normal person and takes 5 mg prednisone per day as maintenance treatment.

Discussion

We reported a young man who suffered from lower limb stiffness with recessive onset and progressive aggravation. He had no previous special or family history. Physical examination found physical signs of upper motor neuron damage. The physical signs showed that the injury position should be above the cervical enlargement, with pyramidal tracts involved. Sensory and ex-

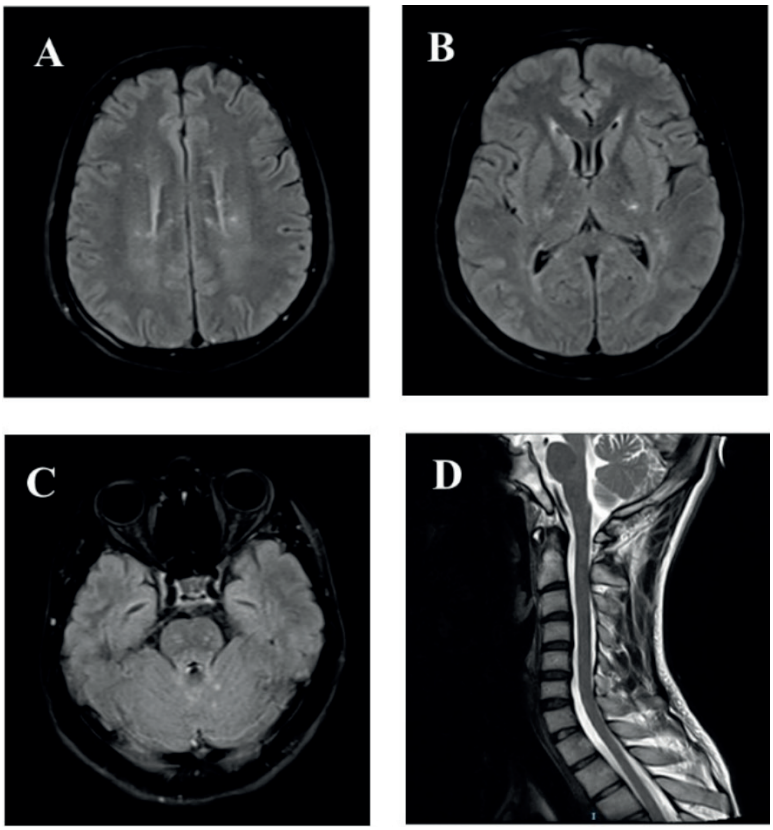

Figure 1. The patient's brain and cervical T2 MRI sequence. Hyperintense signals in the lateral ventricle, brainstem and cerebellum **(A-C)** and slight protrusion of cervical 3-4, 4-5, 5-6 intervertebral discs **(D)**.

trapyramidal systems were not involved. Imaging findings supported lesion localization. Neurodegenerative diseases, such as hereditary spastic paraplegia or motor neuron disease might be initially considered depending on the patient's medical history and physical examination. However, the patient's negative family history and his abnormal brain imaging did not correspond with hereditary spastic paraplegia. Meanwhile, the patient did not have symptoms and physical signs related to lower motor neuron lesions, and therefore, there was no support for the diagnosis of motor neuron disease. Brain MRI showed multiple abnormal signals in the T2 sequence and were identified as inflammation or demyelination lesions. CSF results did not support intracranial infection. Demyelination could not be excluded by negative results of OB and AQP4 antibodies in the CSF. In a blood biochemical examination, anti-ds DNA antibody, anti-Sm antibody and anti-RNP antibody were positive, which suggested SLE. However, the patient did not have skin damage, oral ulcers, arthralgia, kidney injury or haematological system damage throughout the course of the disease. Therefore, NPLE characterized by neurological symptoms was diagnosed. Depending on the classification of NPLE from the ACR, SLE-related demyelination syndrome was considered. The patient had a good treatment response to steroids, which supported the diagnosis of NPLE.

Secondary demyelination is a rare manifestation of SLE with a reported rate of 0.9-2.7%[6,7]. However, demyelination can be the first symptom of SLE in some patients, and the proportion was reported to be up to 40.0%[3]. Most NPLE patients who had demyelination syndrome as the first manifestation presented other SLE manifestations, such as malar rash, oral ulcers, photosensitivity, arthralgia, arthritis, haemolytic anaemia, leukopenia and thrombocytopenia, during 0.5- to 15-year periods of follow-up. Fewer than 10% of patients had no specified SLE manifestations[3]. Our patient did not present SLE manifestations except for neurological symptoms during more than 1 year of follow-up. Secondary demyelination syndrome in SLE can present as NMO, NMOSD, demyelinating syndrome prominently affecting the brainstem, demyelinating syndrome prominently affecting the brain, and clinically isolated syndrome[3,4]. Other common neuropsychiatric symptoms include headache, seizure, aseptic meningitis, cognitive dysfunction and psychosis[8]. Demyelination syndrome with the appearance of spastic paraplegia has not been reported to date.

The relatively symmetrical damage in the pyramidal tracts could explain the clinical manifestations of the patient. The response to steroid therapy also supported the diagnosis of NPLE. Therefore, for patients with clinical manifestations of central demyelination lesions, we should consider the possibility of NPLE. Serological tests and brain MRI should be taken in a timely manner to make an early diagnosis and provide treatment.

Conclusions

This is the first case of spastic paraplegia as the only manifestation in NPLE. Physicians should be aware of this rare manifestation in SLE.

Conflicts of Interest

We acknowledge that there are no conflicts of interest related to this case report.

Consent to Participate

The patient gave the permission to use his clinical image and data.

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