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Case Report

Functional Improvement after Taking Rehabilitation Program in Cerebellar Ataxia in a Patient with Systemic Lupus Erythematosus: a Case Report

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HIGHLIGHTS

- The first report of a patient with SLE and cerebellar atrophy after rehabilitation.
- Cerebellar ataxia is uncommon in SLE patients, and cerebellar atrophy is even rarer.
- BBS, MBI and ICARS improved after intensive rehabilitation treatment.

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Conflict of Interest
The authors have no potential conflicts of interest to disclose.

ABSTRACT

Cerebellar involvement in systemic lupus erythematosus (SLE) is rare, occurring in less than 2% of cases, with cerebellar atrophy on brain imaging even rarer. We describe a case of cerebellar atrophy in a patient with a history of SLE who underwent an intensive rehabilitation program and achieved functional improvement. A 26-years-old female with SLE felt difficult in balancing as well as dizziness and she took magnetic resonance imaging which showed cerebellar atrophy and positron emission tomography-computed tomography which showed markedly decreased signal intensity at bilateral temporal, parietal lobes, cerebellum and moderately at bilateral frontal lobes. A neurological examination showed bilateral nystagmus, intention tremor, bilateral dysmetria, and bilateral dysdiadochokinesia. Prior to the rehabilitation program, the patient's scores on the Berg Balance Scale (BBS) and Modified Barthel Index (MBI) were 4/56 and 37/100, respectively. The patient was hospitalized twice for rehabilitation treatment, which consisted of physical therapy, occupational therapy, therapeutic pool therapy, and robotic-assisted gait training. At discharge, the patient's BBS score was 9/56, and her MBI score was 46/100, and she was able to walk more than 20 m using an anterior walker. In the final 2 weeks before discharge, the patient was trained in the use of an electric wheelchair for outdoor activity.

Keywords: Cerebellar ataxia; Neurological rehabilitation; Systemic lupus erythematosus

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease which is characterized by immunologic abnormalities. SLE can affect any organ of the body including the brain, resulting in neuropsychiatric manifestations. In cases of SLE, 50%–70% of patients have neuropsychiatric manifestations, with cerebellar involvement occurring in less than 2% of cases [1]. In the present report, we describe functional improvements following an intensive rehabilitation program in a 26-years-old female with SLE who developed cerebellar ataxia and cerebellar atrophy.

CASE REPORT

A 26-years-old female was diagnosed with SLE based on immunologic abnormalities, such as polyarthralgia, oral ulcers, photosensitivity, arthritis, thrombocytopenia, leukopenia, and hair loss, in addition to a positive blood test for lupus anticoagulant, antinuclear antibodies, anti-dsDNA, anti-U1-RNP antibody, and anti-Sm antibody. After receiving a diagnosis of SLE, the patient was prescribed oral hydroxychloroquine (200 mg/d) and oral prednisolone (2.5 mg/d).

Eight years later, the patient experienced difficulties with balance, as well as dizziness. Magnetic resonance imaging (MRI) revealed cerebellar atrophy (Fig. 1). In addition, brain fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography (PET-CT) showed markedly decreased signal intensity at the bilateral temporal and parietal lobes and cerebellum and moderately decreased signal intensity at the bilateral frontal lobes (Fig. 2). Various evaluations including blood test and cerebrospinal fluid test revealed no definitive cause of these signs and symptoms other than SLE. Treatment with high-dose corticosteroid therapy for eight days led to partial recovery of the neurological deficit but she still could not stand due to ataxic pattern and felt dizziness after sitting more than 30 minutes. As high-dose corticosteroid therapy was thought to be refractory, treatment with intravenous immunoglobulin (IVIG) and rituximab immunosuppressive was performed but was not effective.

About 11 months after acute phase treatment for neurological symptoms, the patient presented to our hospital and was admitted twice for intensive rehabilitation treatment (1 month each time). A neurological examination performed on the first inpatient day showed bilateral nystagmus and intention tremor. When performing the finger-to-nose test, both sides showed a severe dysmetria pattern, and the rapid alternating movement test revealed dysdiadokokinesia on both sides. The Romberg test and tandem gait test were not performed due to poor standing balance. The patient's International Co-operative Ataxia Rating Scale (ICARS) score was 67/100 in total. In detail, the score was 28/34 for posture and gait disturbance, 32/52 for kinetic functions, 4/8 for speech disorders, and 3/6 for occulomotor disorders. On a cerebral function test (Mini-Mental Status Examination),

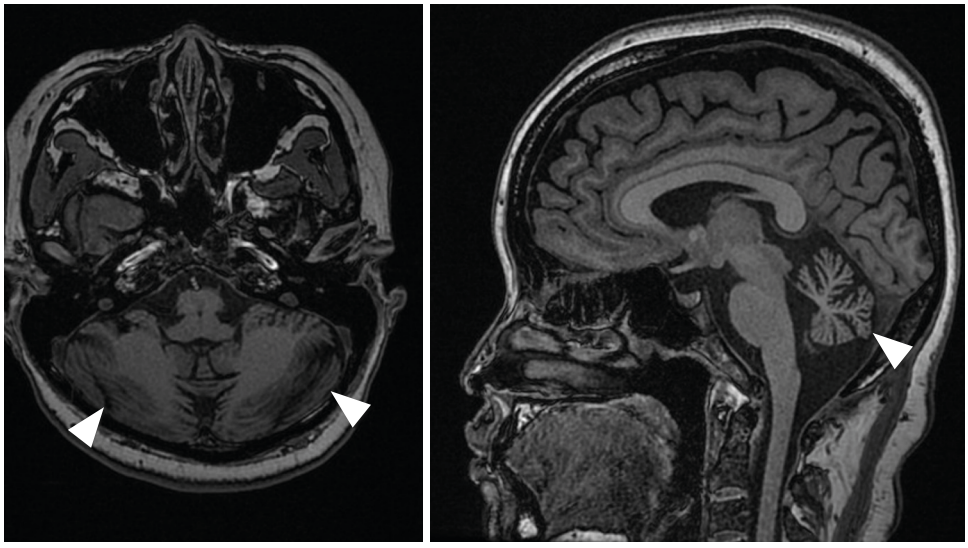

Fig. 1. (Left) Brain MRI (axial view) shows bilateral cerebellar atrophy (arrowhead). (Right) Brain MRI (sagittal view) shows bilateral cerebellar atrophy (arrowhead).

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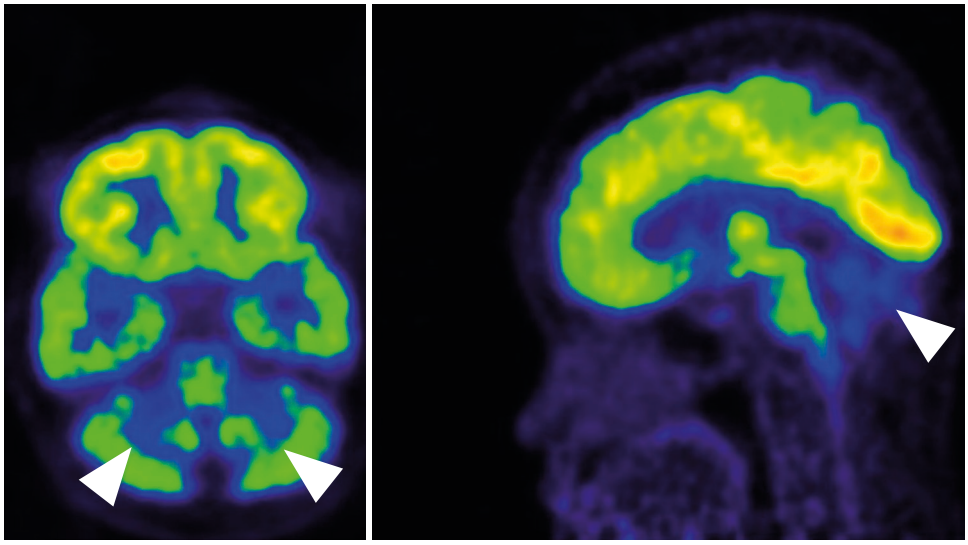

Fig. 2. (Left) Brain PET-CT (axial view) shows markedly decreased signal intensity at the bilateral temporal and parietal lobes and cerebellum and moderately decreased signal intensity at the bilateral frontal lobes. The two arrowheads indicate an area of decreased signal intensity in the cerebellum.
(Right) Brain PET-CT (sagittal view) shows markedly decreased signal intensity at the bilateral temporal and parietal lobes and cerebellum and moderately decreased signal intensity at the bilateral frontal lobes. The arrowhead indicates the area of decreased signal intensity in the cerebellum.

the patient scored 30 points, and all four limbs were classified as grade IV according to the Medical Research Council scale in a manual muscle test of the upper and lower extremities. A sensory examination showed no hypoesthesia or paresthesia of the bilateral upper and lower extremities and trunk. A reflex test revealed a normo-reflex in the bilateral upper and lower extremities. The patient had no visual symptoms, headaches, nausea, vomiting, sphincteric disturbances, weakness, or other neurological symptoms. The patient's Berg Balance Scale (BBS) and Modified Barthel Index (MBI) scores were 4/56 and 37/100, respectively.

During the 2 hospitalization periods, the patient took part in a rehabilitation program comprising physical therapy, occupational therapy, whole body therapeutic pool therapy, and robotic-assisted gait training 5 times a week, 30 minutes each time. The program commenced with sitting balance training, followed by standing balance training and finally gait training using an anterior walker. The patient also performed Frenkel's exercise, including a series of slow, repetitious motions that were performed in different positions when lying down, sitting and standing, 30 minutes at a time and twice a day. Robot-assisted gait training was performed using an end-effector type gait rehabilitation robot system.

At discharge, the patient's BBS and MBI scores were 9/56 and 46/100, respectively. The patient's ICARS score was 59/100 in total. In detail, the score was 26/34 for posture and gait disturbance, 28/52 for kinetic functions, 3/8 for speech disorders, and 2/6 for occulomotor disorders. The patient was able to walk more than 20 m using an anterior walker. As the results of the patient's hand power test and Jebsen–Taylor hand function test were within the normal range, the patient was trained in the use of an electric wheelchair for outdoor activity during the last 2 weeks before discharge.

DISCUSSION

In patients with SLE, cerebellar ataxia is not common and patients presenting with cerebellar atrophy are even rarer [2-8]. The pathogenesis of acute ataxia in lupus may comprise focal involvement or diffuse cerebellar involvement. The former appears in the form of vaso-occlusion or thrombosis, and the latter is due to anti-dsDNA antibodies, cross-reacting with brain anti-N-methyl-D-aspartate receptors, which mediate cerebellar injury or dysfunction. In the present case, no vaso-occlusion or thrombosis was detected on MRI, and brain PET-CT showed markedly decreased signal intensity in a diffuse area involving the bilateral cerebellum. Thus, diffuse cerebellar involvement due to the presence of anti-dsDNA antibodies may be a possible mechanism underlying ataxia in the present case [9].

In a recent cohort study of 325 SLE patients, the authors detected SLE-related autoantibodies (anti-dsDNA, anti-SSA, anti-SSB, anti-RNP, and anti-Sm) in serum [10]. In the same study, neither the total number of autoantibodies nor individual autoantibodies was associated with inflammatory-like lesions or other brain-MRI abnormalities. However, lupus anticoagulant was associated with lacunar infarcts in white matter and brain atrophy [10].

High-dose corticosteroid therapy can play an important role in improving neurological symptoms of patients with cerebellar atrophy [7,9]. However, high-dose corticosteroid therapy may not provide significant functional improvement in some cases [2,3,5]. IVIGs may be used in such cases, which are not responding to conventional immunosuppression including corticosteroid pulses. In neuropsychiatric SLE patients, IVIGs have been successfully used to treat with a broad spectrum of symptoms, including central nervous system manifestations [11]. However, if a patient shows lack of functional outcome despite having completed all such medical treatments, an intensive rehabilitation program focused on balance training and gait training using an appropriate device may lead to functional improvements in patients.

In conclusion, 2 months of intensive rehabilitation treatment resulted in functional improvements in an SLE patient with cerebellar atrophy.

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