Systemic lupus erythematosus (SLE) is a chronic autoimmune disease. Neuropsychiatric systemic lupus erythematosus (NPSLE) is a subtype of SLE specifically involved in neurologic manifestations usually observed in central, peripheral, and autonomic nervous systems and sometimes with psychiatric disorders. Movement disorders such as chorea, ataxia, and hemiballismus have been described in the American College of Rheumatology classification of NPSLE. However, parkinsonism, as a movement disorder, is an extremely rare manifestation of NPSLE. Here, we report a patient of NPSLE with parkinsonism.

**CASE REPORT**

A 17-year-old girl was admitted to our hospital after one month of fever, hand erythema, and arthralgia. There were no significant findings in her personal or family history. Results from blood test showed leucopenia (white blood cell 2.7*10^9/L), positive antinuclear antibody (1:1000), anti-double stranded (ds) DNA, anti-Sjögren syndrome A antigen, anti-Sjögren syndrome B antigen, and anti-ribosome P protein; negative antiphospholipid antibody, anti-beta2GP1 antibody, and anti-neutrophil cytoplasmic antibodies (low complement 3 (0.31 g/L) and high erythrocyte sedimentation rate (89 mm/h). Urine test showed no hematuria or proteinuria. According to the 1982 revised criteria for the classification of SLE, she was diagnosed with SLE. The Systemic Lupus Erythematosus Disease Activity Index score was 16. She was administered with intravenous methylprednisolone pulse therapy, intravenous cyclophosphamide, and intrathecal injection of methotrexate plus dexamethasone were prescribed. Two months later, the patient returned with complete clinical recovery of neuropsychiatric symptoms and signs.

**Keywords:** Cyclophosphamide; glucocorticoid; methotrexate; neuropsychiatric systemic lupus erythematosus; parkinsonism.
during this one-week period, she developed a progressive slowdown of motor response, movement, thinking, speech, and reaction to stimuli. She did not complain of any limb weakness or numbness, giddiness, diplopia, tinnitus, urinary or bowel dysfunction. Neurological examinations showed that she was fully oriented with no evidence of cognitive dysfunction, aphasia, paresthesia, meningism, lateralizing neurological deficits, or cranial nerve palsies. With disease progression, she developed obvious expressionless face, bradykinesia, marked rigidity, and hypermyotonia in her extremities. She also had a festinating gait with only slight arm swing, and her gaits turned and started in hesitation but without rest or action tremor.

Head magnetic resonance imaging showed symmetrical long T1 signal, slight long T2 signal, and high T2 flair signal in bilateral basal ganglia, hippocampus, insular lobe, and subcortex of frontal gyrus rectus. Bilateral external capsule showed long T1 signal, long T2 signal, and high T2 flair signal (Figure 1). Enhanced magnetic resonance imaging showed a slightly increased signal intensity of bilateral basal ganglia. Lumbar puncture showed that the cerebrospinal fluid (CSF) was colorless and transparent with CSF pressure at 120 mmH2O. Biochemical test of CSF showed normal levels of protein (175 mg/L), glucose (3.1 mmol/L), lactate dehydrogenase, chloride, and adenosine deaminase, but a slight elevation of white blood cells (25*10^6/L). CSF smear, culture, Gram staining, acid-fast staining, tuberculosis DNA, virus antibody test, and fungal test were all negative. Autoantibodies to dsDNA and ribosomal P protein were positive in the CSF, but anti-phospholipid, anti-beta2GP1, anti-neuronal, anti-glial fibrillary acidic protein, anti-endothelial cell, and anti-N-methyl-D-aspartate antibodies were negative. Since none of the drugs administered appeared to have potential side effect of parkinsonism and no other causes of parkinsonism could be recognized for this particular patient, she was considered to have NPSLE.

High dose intravenous methylprednisolone (500 mg/day) was prescribed for three days followed by oral prednisolone (1 mg/kg/day), and intravenous cyclophosphamide (600 mg/m² body surface

![Figure 1. (a-c) Magnetic resonance imaging of the head showed symmetrical lesions in bilateral basal ganglia, hippocampus, insular lobe, and subcortex of frontal gyrus rectus. (d-f) Magnetic resonance imaging of the head was normal at patient’s nine-month follow-up.]
area) was prescribed once. Intrathecal injection of methotrexate (MTX) (10 mg) and dexamethasone (DXM) (10 mg) were applied once a week for two weeks. After these treatments, the patient showed steady improvements in her gait, speech and movements. Two weeks later, her blood showed normal level of complement 3 (0.84 g/L) and erythrocyte sedimentation rate (18 mm/h). She was discharged on maintenance of oral prednisolone (30 mg/day), oral hydroxychloroquine (200 mg/day), and intravenous cyclophosphamide (800 mg/month). At the patient’s two-month follow-up visit, we noticed that she had normal facial expression, speech, movement, and gait. Complement 3 (0.86 g/L) and erythrocyte sedimentation rate (12 mm/h) remained normal and the titer of antinuclear antibody decreased to 1:40. The Systemic Lupus Erythematosus Disease Activity Index was reevaluated with a score of 0. Patient was still in remission at nine-month follow-up, and her head magnetic resonance imaging was normal (Figure 1).

**DISCUSSION**

Neuropsychiatric systemic lupus erythematosus associated parkinsonism was first described by Seminario and Pesano in 1930. Since then, only about 30 NPSLE cases with parkinsonism have been reported to date (Table 1). Autoantibodies and immune-mediated vasculopathy was suggested in the pathogenesis of parkinsonism in NPSLE. Kunas et al. found antibodies to dopaminergic cells in serum from the NPSLE patients manifested with parkinsonism. They concluded that anti-dopamine antibodies could be specific to NPSLE patients with parkinsonism. However, to our knowledge, there are no controlled trials of any treatment for NPSLE with parkinsonism. Improvements have been reported in those patients treated with combination of steroids and cyclophosphamide, with or without antiparkinsonian drugs. In two cases, intravenous injection of immunoglobulins appeared to be effective after methylprednisolone and cyclophosphamide failed. Intrathecal injection of MTX plus DXM, which was assumed to enhance...
the immunosuppressive effects but reduce side effects, has been shown to improve the outcomes of NPSLE patients without parkinsonism. To our knowledge, our patient was the first NPSLE patient with parkinsonism reported to accept intrathecal injection of MTX plus DXM and obtain a satisfactory outcome. Although there was no control in our therapeutic regimen, results from this particular case support the notion that intrathecal injection of MTX and DXM, plus high dose intravenous methylprednisolone and cyclophosphamide for NPSLE patients with parkinsonism may be a plausible therapeutic strategy.

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