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Neuromyelitis Optica Spectrum Disorder Presenting with Pseudoathetosis

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Dear Editor,

Pseudoathetosis is a movement disorder that is characterized by involuntary, slow, and writhing movements. It can develop from lesions at all levels of proprioceptive sensory pathways, including the posterior column of the spinal cord, parietal cortex, thalamus, and peripheral nerves.[1,2] Various conditions that involve the posterior column have been reported to cause spinal pseudoathetosis, including multiple sclerosis, myelitis, spinal cord infarct, trauma, tumors, spondylotic myelopathy, leprosy, and vitamin B12 deficiency.[2] However, neuromyelitis optica spectrum disorder (NMOSD), which is an acquired immune-mediated inflammatory disease of the central nervous system (CNS) characterized by the presence of an antibody to aquaporin-4,[3,4] has not been reported previously as a cause of pseudoathetosis. We report a case that was diagnosed as pseudoathetosis caused by cervical myelitis associated with an aquaporin-4 antibody.

A 69-year-old woman who was otherwise healthy noticed abnormal movements in her hands. The initial diagnosis at the outpatient clinic was dystonic chorea. The abnormal movements progressed to the point where, she found it difficult to handle small objects with either hand. Right leg weakness was noted 1week later. On admission, her general status was unremarkable. Cranial nerve examination produced normal finding. Medical Research Council (MRC) grade 2 weakness was present in the right leg, but her deep tendon reflexes were normal. Plantar responses were flexor bilaterally. A sensory examination revealed severe reduction in the joint position and vibration sense in both the hands and feet. However, the touch, pain, and temperature senses were normal. Her fingers exhibited uncontrolled postures and movements. The slow and involuntary movements were more obvious when she closed her eyes (Supplementary Video 1 in the online-only Data Supplement). Spinal magnetic resonance imaging (MRI) revealed a hyperintense posterior column lesion in T2-weighted images of the spinal cord that extended from levels C1 to C8 (Fig. 1). No lesion suggestive of multiple sclerosis was observed on brain MRI.

The findings of blood tests including thyroid, vitamin B12, folic acid, anti-dsDNA antibodies, rheumatoid factor, and VDRL/TPHA were negative. The cerebrospinal fluid (CSF) examination showed slightly elevated proteins (95.7 mg/dL), and oligoclonal bands were found. We also applied a serologic test for an aquaporin-4 antibody to differentiate NMOSD from the other causes of longitudinally extensive transverse myelitis, which detected the antibody. Although visual evoked potentials were not investigated due to the patient's refusal, she had no visual symptoms and had no prior history suggestive of optic neuritis.

The patient was treated with high-dose methylprednisolone (1,000 mg/day intravenously) for 5 days. Follow-up MRI performed 1 week after the steroid pulse therapy showed a decreased extent of the initial T2-weighted hyperintense lesion involving the posterior columns of the cervical spine. The abnormal postures and movements of her fingers had completely disappeared at the 3-month follow-up, suggesting reversibility, and her right leg weakness

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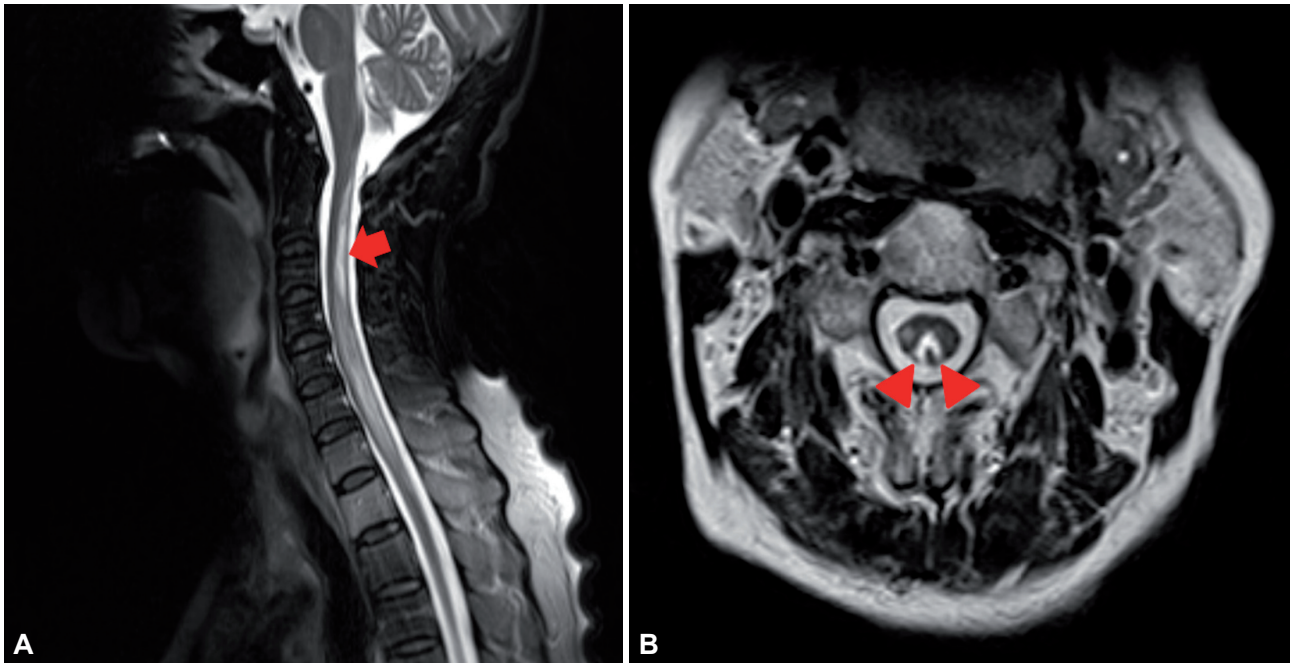

Fig. 1. Spinal cord MRI. A: Sagittal T2-weighted image shows thickening of the cord from C1 to C8 with patchy areas of intraparenchymal hyperintensity (arrow). B: Axial T2-weighted image reveals bilateral involvement of the posterior spinal cord in the upper cervical region (arrowheads).

had improved to MRC grade 4. The vibration and position sensory impairments had been ameliorated. At the last follow-up performed 18 months after her initial attack, she had experienced no relapse while receiving treatment with prednisolone at 5 mg every other day.

Our patient met the 2015 revised diagnostic criteria of NMOSD[3] by having one core clinical characteristic (acute myelitis) and positivity in a test for an aquaporin-4 antibody. Although the presence of CSF oligoclonal bands was a red flag for NMOSD in this case, its diagnostic sensitivity and specificity are modest because oligoclonal bands can be seen in as many as 20% of patients with NMOSD.[3]

There are some initial clinical presentations that are characteristic of NMOSD but not specific to this disease,[3] include severe, bilateral simultaneous or rapidly sequential optic neuritis and longitudinally extensive transverse myelitis. Painful tonic spasm associated with myelitis reportedly occurs in 17.2% to 25% of NMOSD patients, but other involuntary movements have been reported only rarely.[4] To the best of our knowledge, this is the first report of pseudoathetosis caused by NMOSD. Pseudoathetosis related to CNS demyelination has been reported only in some cases of multiple sclerosis.[2,5,6] Unlike multiple sclerosis, NMOSD cord lesions typically involve the central gray matter, with lesions involving predominantly peripheral white matter such as the posterior columns being uncommon in NMOSD.[3] However, a recent study that examined the pathology of spinal cord lesions from 11 autopsied NMOSD cases found that posterior and lateral columns of the white matter were frequently affected in addition to the central gray matter.[7] The present case suggests that pseudoathetosis can occur as the presenting symptom of NMOSD, and thus it should be considered in the differential diagnosis of pseudoathetosis, as well as multiple sclerosis.

Supplementary Video Legend

Video 1. Pseudoathetosis in fingers. Abnormal postures of the fingers were noted when the patient's eyes were open. When she was asked to close her eyes, dropping of both arms and involuntary slow movements of the fingers were observed.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10-988/jcn.2018.14.1.123.

Conflicts of Interest

The authors have no financial conflicts of interest.

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