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Single Case – General Neurology

Two Cases of Very-Late-Onset Neuromyelitis Optica Spectrum Disorder (NMOSD) in Patients over the Age of 80

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Keywords
Neuromyelitis optica · Very late onset · Anti-aquaporin-4 antibody · Transverse myelopathy

Abstract
We report two cases of very-late-onset neuromyelitis optica spectrum disorder (NMOSD) in patients over the age of 80 with transverse myelopathy as the initial manifestation. In both cases, the patients presented with paraplegia and sensory, bladder, and rectal disturbances. Thoracic magnetic resonance imaging showed longitudinal high-intensity signals on a T2-weighted image. The patients received high-dose methylprednisolone. Their serum was positive for anti-AQP4 antibody (cell-based assay) during the clinical course. They were diagnosed with NMOSD and treated with immunoadsorption, plasmapheresis, and followed up with daily prednisolone. Very-late-onset NMOSD in patients over the age of 80 has only rarely been reported. The present cases suggest that NMOSD should be considered for elderly patients presenting with transverse myelitis. Early diagnosis and treatment are important.

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Introduction

Neuromyelitis optica (NMO) is a rare inflammatory demyelinating disease of the central nervous system affecting predominantly women, most frequently in their 30s to 40s [1–5]. The anti-aquaporin-4 (AQP4) antibody has been identified as a disease-specific autoantibody in NMO patients [1–5]. NMO and its incomplete forms are referred to as NMO spectrum disorders (NMOSD) [1–5]. Very-late-onset NMOSD in patients over the age of 80 has only rarely been reported. We present two cases of very-late-onset NMOSD in patients over the age of 80 with transverse myelopathy as the initial manifestation.

Case Report

Case 1

An 82-year-old woman was admitted for acute gait disturbance and hypoesthesia over 3 days. A general physical examination was normal. Neurological examination revealed muscle weakness of grade 4/5 affecting the right lower limb and grade 1/5 affecting the left lower limb. Deep tendon reflexes were hyperactive in both limbs. Babinski and Chaddock signs were positive on both sides. There was a loss of position and vibration sense combined with hypesthesia below Th10. She had bladder and rectal disturbance. Her Expanded Disability Status Scale (EDSS) score was 8.0. Findings from routine serum studies were normal. Cerebrospinal fluid (CSF) analysis showed a cell count of 2/mL (100% mononuclear cells), normal glucose level, an increased protein concentration of 48 mg/dL, a myelin basic protein (MBP) level of 862 mg/dL (normal range <102 mg/dL), immunoglobulin G (IgG) and albumin ratios of 1.18 (normal range <0.7), and positive oligoclonal bands. Spinal magnetic resonance imaging (MRI) on a T2-weighted image (T2WI) demonstrated two high signals extending from Th1 to Th2 and Th7 to Th9 in the central part of the cord (Fig. 1a–d, arrows). The lesions showed no contrast enhancement. Brain MRI showed no abnormality. As autoimmune myelitis was suspected, she received high-dose methylprednisolone (1,000 mg/day for 3 days, 3 courses). Her serum was positive for anti-AQP4 antibody (cell-based assay) 17 days after the onset. She was diagnosed with NMOSD and treated with immunoadsorption plasmapheresis and followed up with daily prednisolone (30 mg). Her condition improved and she was able to walk using a walking instrument. Repeat MRI showed marked regression of thoracic lesions 47 days after the onset. She was discharged to another hospital for rehabilitation 50 days after the onset with daily prednisolone (15 mg). Her EDSS score was 5.5.

Case 2

An 80-year-old man was admitted for subacute gait disturbance. His bilateral lower limb impairment had developed over 2 weeks. A general physical examination was normal. Neurological examination revealed muscle weakness of grade 4/5 affecting both lower limbs. Deep tendon reflexes were absent in both limbs without pathological reflexes. There was loss of deep sensation below T10. He had bladder and rectal disturbance. His EDSS score was 8.5. Findings from routine serum and CSF studies were normal. Spinal MRI on T2WI revealed high signals extending from Th9 to Th12 in the central part of the cord (Fig. 1e, f arrow). The lesions showed no contrast enhancement. Brain MRI showed no abnormality. As autoimmune myelitis was suspected, he received high-dose methylprednisolone (1,000 mg/day for 3 days, 2 courses). His serum was positive for anti-AQP4 antibody on the 15th day of his hospitalization. He was diagnosed with NMOSD and treated with immunoadsorption plasmapheresis and

followed up with daily prednisolone (30 mg). The muscle strength of the lower limb slightly improved; however, he was bedridden, and the paraplegia persisted. Repeat MRI showed marked regression of thoracic lesions on the 43rd day of his hospitalization. He was discharged to another hospital for rehabilitation 50 days after his hospitalization with daily prednisolone (10 mg). His EDSS score was 7.5.

Discussion

We report 2 cases of very-late-onset NMOSD in patients over the age of 80 with transverse myelopathy as the initial manifestation. NMOSD is an inflammatory demyelinating disease of the central nervous system affecting predominantly women, most frequently in their 30s to 40s [1–5]. NMOSD has been described in elderly patients [6–9]; however, very-late-onset NMOSD in patients over the age of 80 is extremely rare. Late-onset NMOSD is a severe disease with a high rate of motor impairment and death [6–9]. In addition, its onset in elderly people is characterized by increased risks of disability, poor response to treatment, and many cases of death due to infection or opportunistic infection during the course [6–9]. The patients are also characterized by a larger representation of initial myelitis and fewer brain MRI lesions [6–9]. The same tendency was observed in our cases. NMOSD was considered as the first-line diagnosis because of longitudinal spinal cord lesions in our cases. However, vascular, infectious, and orthopedic myelopathy are the most common causes of myelopathy in elderly people. Delayed diagnosis and treatment could lead to an unfavorable outcome. Early diagnosis and treatment are important, and NMOSD should be considered in elderly patients presenting with transverse myelitis.

Statement of Ethics

The patients gave informed consent for the publication of this case report.

Disclosure Statement

The authors state that they have no conflicts of interest.

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Author Contributions

Shunya Fujiwara and Yasuhiro Manabe designed the case report and wrote the manuscript. Shunya Fujiwara, Yasuhiro Manabe, Ryuta Morihara, Taijun Yunoki, Syoichiro Kono, and Hisashi Narai contributed to the diagnosis, physical examination, and testing of the patient. Koji Abe supervised this case report and clinical practice. All authors read and approved the final manuscript.

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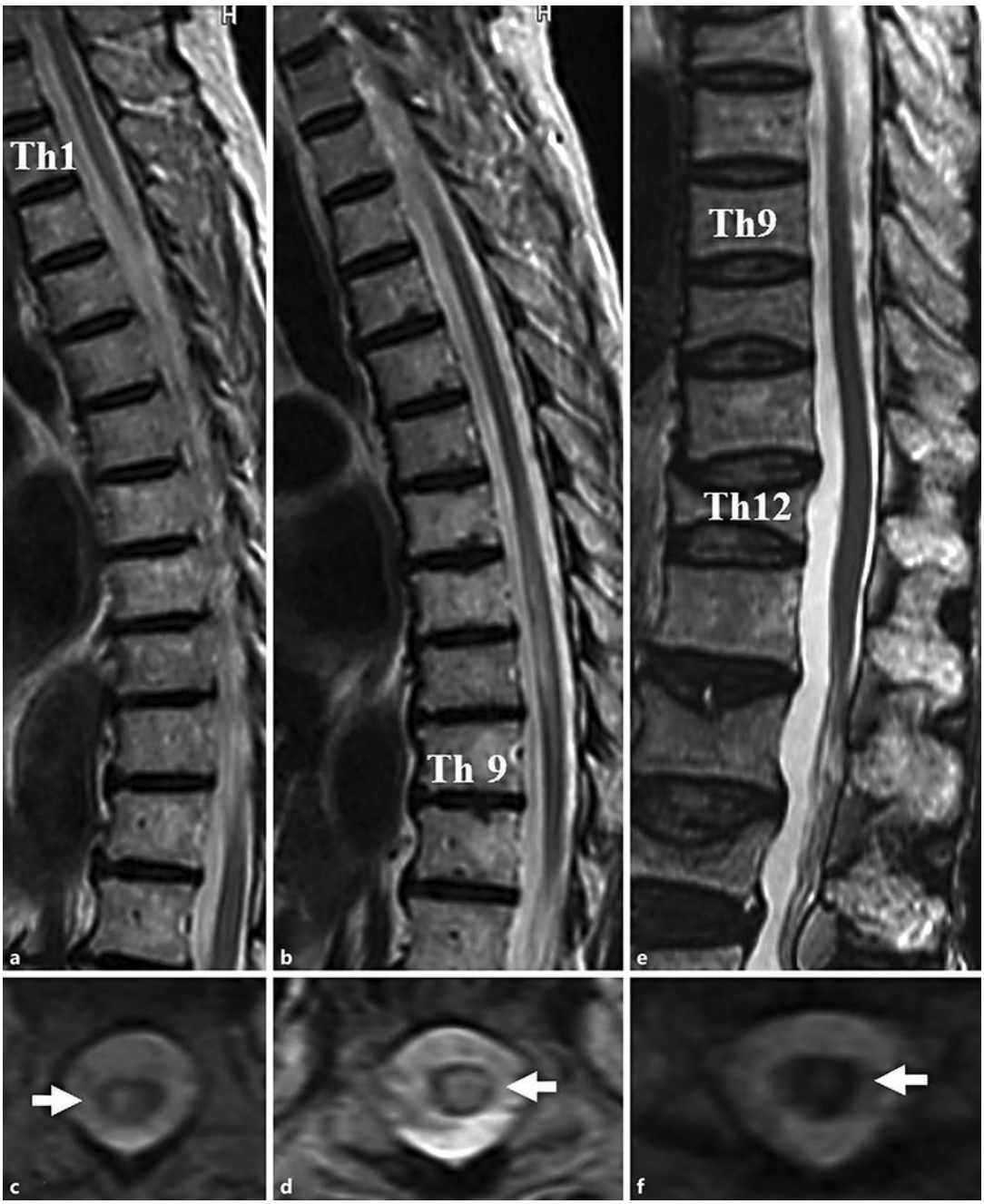

Fig. 1. Sagittal thoracic spine magnetic resonance imaging (MRI) on admission in case 1 showing high-intensity lesions extending from Th1 to Th2 and from Th7 to Th9 on T2-weighted image (T2WI) (**a**, **b**). Axial thoracic spine MRI showing high signals in the central part of the cord at the Th2 and Th8 levels on T2WI (**c**, **d**, arrows, respectively). Sagittal thoracic spine MRI on admission in case 2 showing high signals extending from Th9 to Th12 on T2WI (**e**). Axial thoracic spine MRI showing high signals in the central part of the cord at the Th9 level on T2WI (**f**, arrow).