Case Report

Very Late-Onset Neuromyelitis Optica Spectrum Disorder Presenting with Falls and Monoparesis: A Case Report

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Here, we report a case of very late-onset neuromyelitis optica spectrum disorder [NMOSD] in an 83-year-old female who presented with monoparesis at right leg and subsequent falling that resulted in right femoral neck fracture. The patient was initially diagnosed as having lumbar spondylosis with radiculomyelopathy. As the disease progressed, she developed quadriparesis that resulted from extensive transverse cervical myelitis with positive anti-aquaporin-4 antibody. This finding helped to guide us to a diagnosis of NMOSD. Accordingly, any patient, regardless of age, that presents with extensive transverse myelitis should be considered as NMOSD.

Keywords: Neuromyelitis optica, Late-onset, Elderly

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Neuromyelitis optica [NMO] is an inflammatory demyelinating disease that primarily affects the spinal cord and optic nerves[1]. In Western countries, NMO is most prevalent in the third to fourth decades of life as it has been shown in Asian countries[2,3]. In the elderly, NMO can present without optic neuritis and is referred to NMO spectrum disorder [NMOSD][4]. Anti-aquaporin-4 antibody is helpful in diagnosis of NMOSD with high sensitivity and specificity[5]. However, older patients might be misdiagnosed as having lumbar spondylosis at initial presentation. This potential for misdiagnosis and the resulting delayed diagnosis and treatment of NMOSD may adversely affect patient outcome.

Case Report

An 83-year-old woman was admitted to our center with a right-side femoral neck fracture that had been sustained in a fall. Two weeks before being admitted, she developed progressive weakness with tingling sensation in her right leg and coexisting new onset constipation and urinary retention. Magnetic resonance imaging [MRI] of lumbar spine revealed severe lumbar spondylosis with grade II spondylolisthesis at L4-L5, with bilateral L4 nerve root compression. Lumbar spondylosis with radiculomyelopathy was diagnosed and medications to control her symptoms were prescribed.

Her vital signs at admission were, temperature 36.0°C, heart rate 62 bpm, respiratory rate 18 breaths/minute, and blood pressure of 129/74 mmHg. Neurological examination found the patient to be alert and cooperative. She had normal cranial nerve examination, muscle tone, proprioception, and deep tendon reflexes. Muscle power was decreased at right lower extremity at grade 3/5, with some limitation in evaluation due to pain. Pinprick sensation was reduced at the right leg. Babinski reflex was present bilaterally.

The patient underwent uneventful bipolar hemiarthroplasty on the second day of admission. She subsequently developed intractable hiccups with no observable abnormality in metabolic profiles. Hiccups were slightly improved after metoclopramide administration. On the fifth day of admission, our patient developed progressive weakness in her left upper and both lower extremities, which was followed shortly thereafter by progressive weakness in her right upper extremity. She also suffered from painful muscle spasms in her left arm and both legs. Physical examination revealed increased muscle tone at left upper and both lower extremities with hyperreflexia. Motor power of her left upper extremity was 3/5 and both lower extremities were 2/5, while her right upper extremity was 4/5. Progression of impairment in pinprick sensation was also observed, with left side

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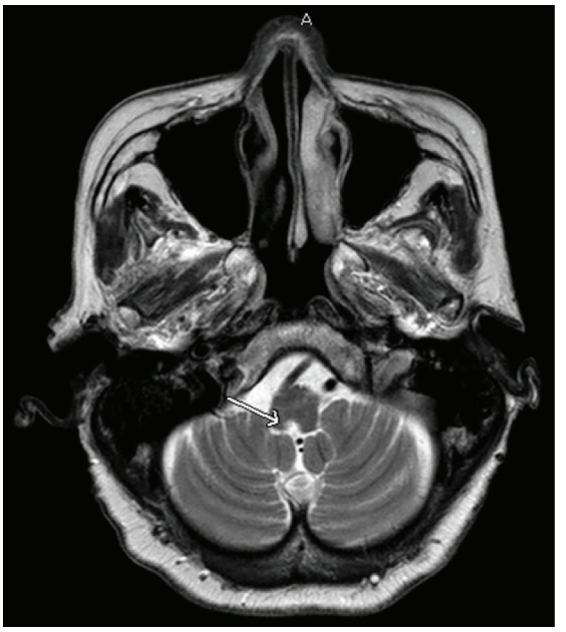

Figure 1. MRI of the brain, T2-weighted image, showing axial view of brain showing hypersignal at the right posterior aspect of medulla.

from C3 level downward and right side from L4 level downward. Proprioception was impaired at left upper and both lower extremities. Eye examination revealed no evidence of optic neuritis.

MRI of the brain and spine revealed multiple small high-intensity lesions along the ependymal lining at the bilateral lateral ventricles and at the right posterior aspect of the medulla with some areas of linear and nodular periventricular enhancement, multiple levels of intramedullary hypersignal on T2-weighted along the cervicothoracic level, including upper C2 to upper C4, lower C5 with peripheral enhancement, at T1 level predominated at the right side and the center of this cord level with peripheral enhancement, and from T7 to lower T9 level (Figure 1-3).

Cerebrospinal fluid examination showed a cell count of 17/mm3 (100% lymphocyte), protein concentration of 50 mg/dL, normal glucose level, no abnormal cells, and open pressure was 10 cmH_2O. Viral serologies were all negative. Results of anti-neuronal antibody and autoimmune encephalitis panels were both negative. Anti-aquaporin-4 antibody test was performed, which showed a positive result with antibody titers greater than 1:10. NMOSD with anti-aquaporin-4 antibody was diagnosed and the patient was prescribed a 1 g dose of intravenous methylprednisolone every 24 hours for eight days. At the end of that regimen, she was given a 25 mg dose of azathioprine orally every 24 hours together with prednisolone 1 mg/kg/day for two weeks. After treatment, there was no observable improvement in her motor power, but her hiccups and muscle spasm symptoms were both improved. Finally, the patient was discharged to her home with 20 mg/day of prednisolone.

Discussion

This report presents the case of an elderly woman who presented with fall due to acute myelitis and who was first diagnosed with NMOSD at 83 years of age. The average age of NMOSD onset is 30 to 40 years, with a higher prevalence among females[3,6,7]. NMOSD patients were more likely to present with transverse myelitis than optic neuritis[7]. Myelitis is also a common initial presentation in both late-onset patients (older

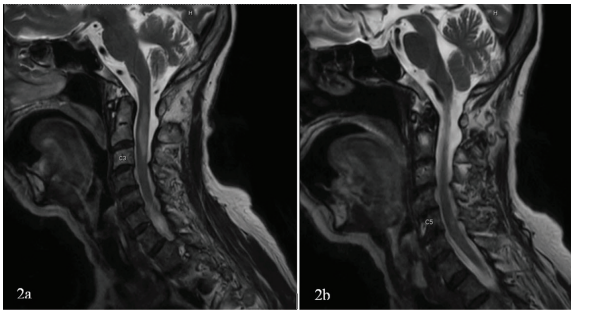

Figure 2. MRI of the cervico-thoracic spine, T2-weighted image, showing: (2a) sagittal view of cervical spine, showing hypersignal lesion along C2 to C4 level; (2b) showing hypersignal lesion at C5 level.

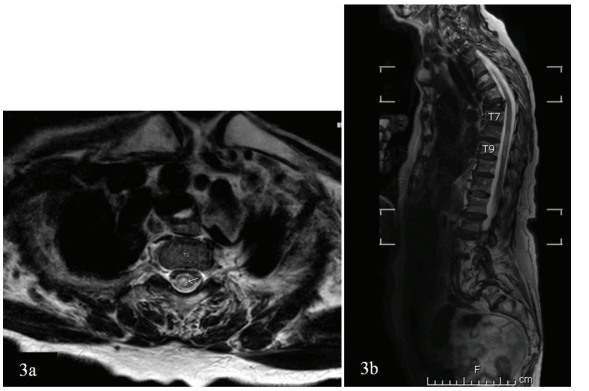

Figure 3. MRI of the cervico-thoracic spine, T2-weighted image, showing: (3a) axial view of thoracic spine, showing hypersignal lesion at T1 level predominated at the right side and the center of the cord level with peripheral enhancement; (3b) sagittal view of thoracic spine, showing hypersignal from T7 to T9 level.

Table 1. Case reports profiling patients with very late-onset NMOSD (age over 75 years)

Country	No./age at onset (year)	Clinical characteristics
France[8]	1/77	Myelitis onset; relapse after 1 year
USA[9]	1/85	Myelitis onset; no long-term follow-up
Germany[4]	1/79	Myelitis onset; several relapses with myelitis and optic neuritis
Germany[4]	1/83	Myelitis onset; relapse-free for 2 years
Germany[4]	1/88	Myelitis onset; death within 1 year
Japan[10]	1/84	Brainstem onset; recurrent brainstem lesions and subsequent severe longitudinally extensive cervical cord lesions
Japan[11]	1/89	Optic neuropathy and myelitis onset
Japan[6]	1/90	Myelitis onset

No. = number of patients

than 50 to 75 years of age)[4] and very late-onset patients (older than 75 years of age) (Table 1).

In some previous case reports, elderly patients presented with NMOSD that mimicked either cervical or lumbar spondylotic myelopathy[4,6]. This potential for delayed diagnosis and treatment of NMOSD could result in subsequent relapses and even death in some patients.

However, our patient initially presented with right monoparesis that was thought to be spondylosis with radiculomyelopathy, but incompatible signs and symptoms were positive for Babinski reflex bilaterally, increase in right lower extremity muscle tone, and history of bowel and bladder symptoms, all of which made spinal cord lesion more likely. Then she developed intractable hiccups and progressive signs of transverse myelitis and muscle spasms, with MRI findings of longitudinally extensive transverse myelitis lesion at the cervical and thoracic spinal cord levels and associated dorsal medulla lesion (area postrema syndrome). These findings provided clinical clues for NMOSD, so anti-aquaporin-4 antibody was requested and NMOSD was diagnosed according to NMOSD diagnostic criteria[12].

Due to possibility that this disorder could be paraneoplastic syndrome (especially in the elderly)[13], cancer screening, autoimmune disease, and antineuronal antibody testing was ordered and all were found to be negative for this patient.

In conclusion, NMOSD is a rare disease among the elderly that has a poor prognosis if not promptly diagnosed and treated. In elderly patients with extensive transverse myelitis, physicians are encouraged to investigate and remain vigilant for the presence of NMOSD.

What is already known on this topic?

NMOSD is an inflammatory demyelinating disease that primarily involves the spinal cord and optic nerves. The average age of NMOSD onset is 30 to 40 years, with a higher prevalence among females. Previous case reports showed NMOSD could be found in the elderly patients even though it was rarely at initial presentation in the patients[4,6,8,9]. In Thailand, there is limited information of NMOSD in very old patients.

What this study adds?

This topic shows the rare case of NMOSD in Thailand. The patient presented with monoparesis and fall that was initially misdiagnosed as having lumbar spondylotic radiculomyelopathy. After the disease progressed, the patient developed quadriparesis that resulted from extensive transverse cervical myelitis with positive anti-aquaporin-4 antibody. Accordingly, NMOSD was diagnosed. The delay diagnosis might affect the patient outcome; therefore, NMOSD should be of concern in the elderly patients with extensive transverse myelitis.

Authors' contributions

Soontrapa P: acquisition and interpretation of data, preparation of manuscript, and the bibliographic search. Srinonprasert V: study concept, and revision of the manuscript. Chotinaiwattarakul W: study concept, and interpretation of data. Suraarunsumrit P: study concept, interpretation of data, bibliographic search, and the preparation and revision of the manuscript.

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Potential conflicts of interest

The authors declare no conflict of interest.

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