Neuromyelitis optica spectrum disorder with delayed lesion on spinal cord magnetic resonance imaging

Ji Eun Han, Hye Rin Yoon, Joong hyun Park, Jeong Yeon Kim*

*Correspondence

Jeong Yeon Kim, MD, PhD

Department of Neurology,
Inje University, Sanggye Paik Hospital
1342 dongil-ro, Nowon-gu, Seoul, Korea
Tel: +82-2-950-1090
jykimmd@paik.ac.kr

Introduction

The term Neuromyelitis optica spectrum disorder (NMOSD) is a recently proposed for neuromyelitis optica and related syndromes. Along with serum anti-aquaporin 4 antibody status, detection of a lesions extending over ≥ 3 contiguous spinal cord segments (longitudinally extensive transverse myelitis, LETM) associated with acute myelitis is the most specific neuroimaging characteristic of NMOSD. [1] We present a case of NMOSD with delayed lesion on spinal cord magnetic resonance imaging.

Case Report

A 36-year-old woman came to our emergency department with a 2-day history of inability to walk and voiding difficulty. She felt a weakness of both legs during stretching, and did not have lower back pain. She had no recent history of infection and vaccination. She reported hospitalization because of a loss of vision in her right eye five years ago. Her visual acuity was 0.1 with reduced color perception, and both pupils showed a poor response to light stimulation. Blood pressure was 130/90 mmHg, pulse was regular with 94 beats per minute and respirations 12 breaths per minute. There were no skin lesions, arthralgia, eyeball pain and photosensitivity. Neurologic examination revealed diminished motor strength in lower extremities with Medical Research Council (MRC) grade 2/2 in hip flexion/extension, knee flexion/extension, ankle dorsiflexion/plantarflexion, and foot inversion. There was hyperesthesia below the T10 level, bilateral positive Babinski sign and ankle clonus, and increased deep tendon reflexes on the knee. No neck rigidity and cranial nerves abnormality was noticed. Inflammatory parameters were not elevated and chest X-ray was normal. Routine laboratory data was normal, including complete blood count (Hemoglobin 14.1 g/dL, white blood cell count 7.190/ μ L with 81% neutrophils), renal, liver and thyroid function tests. Cerebrospinal fluid (CSF) analysis

revealed pleocytosis (lymphocyte count of 50/mm³) with elevated albumin concentration (746 mg/dL; normal range 10-30), but otherwise including CSF glucose was normal. CSF oligoclonal band was analyzed with concurrent serum sample and the results were negative. Visual evoked potential test showed no detection of P100 latency from right eye stimuli. Serum anti-aquaporin-4 (anti-AQP-4) antibody test was positive. To exclude other autoimmune diseases, we evaluated autoimmune antibodies in serum. Rheumatoid factor, c-antineutrophil cytoplasmic antibodies (ANCA), p-ANCA, anti-ds DNA, and paraneoplastic antibodies including anti-Hu, anti-Yo, and anti-Ri were negative results. Magnetic Resonance Imaging (MRI) of the brain and thoracic spinal cord was performed the next day after the hospitalization (after 3 days of symptoms onset) to confirm the presence of other differential diseases. No lesions such as lesions in brainstem, incidental or unlocalizable finding of AVF or white matter changes suspicious CNS demyelinating disease were not found (Figure 1 A, B).

Despite of the normal thoracic spinal MR imaging, presence of clear clinical evidence of thoracic myelopathy and history of optic neuropathy with abnormal evoked potential findings were felt to be more consistent with a neuromyelitis optica or multiple

sclerosis, so she was started on a five day course of intravenous methylprednisolone (1 g IV qd for 5 days and then tapered to 1 mg/Kg/d) was started. Five days later (after 8 days of symptoms onset), repeat T2-weighted thoracic spine MRI (Fig. 1 C, D) revealed markedly hyperintensity lesion in T2-10 spinal cord with moderate swelling of the cord at these levels. The patient was finally diagnosed Anti-aquaporin 4 antibody positive neuromyelitis optica spectrum disorder and she was treated with immunosuppressive treatment (azathioprine 2mg/kg daily). She exhibited slowly improvement of weakness with a comprehensive rehabilitation program, although she remained paraplegic and confined to a wheelchair.

Spinal MR imaging with and without contrast is the initial investigation of choice for evaluation of acute myelopathy. 2 In our patient, the initial clinical manifestations indicated a thoracic level suggesting acute transverse myelitis. But, no abnormal findings were observed in the initial MR imaging, and characteristic longitudinally extensive transverse myelitis findings were detected only in the follow up spinal imaging performed 5 days later. We concluded that there was no abnormality in the initial imaging because lengthy T2-weighted lesion did not developed fully in the few days after clinical symptom onset. Initial negative spinal MR imaging does not exclude neuromyelitis optica spectrum disorder and a follow imaging investigation with closed neurological observation is necessary.

Figure. Brain and spine MRIs of a patient. Axial FLAIR image of brain (A) and sagittal T2- weighted MRI of thoracic spinal cord after 3 days of symptoms onset (B) shows normal brain and spinal cord. Follow up spinal MRI shows longitudinally extensive transverse myelitis (LETM) lesions (red arrows) on thoracic sagittal (C) and axial (D) vertebral segments after 8 days of symptoms onset.

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