Case Report

Neuromyelitis optica with rapid respiratory failure: a case report

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Background: Neuromyelitis optica is an inflammatory demyelinating disease of the central nervous system and is characterized by severe optic neuritis and transverse myelitis.

Case Presentation: The patient was a 74-year-old man with pneumonia. On admission, he exhibited lower limb weakness and rapid respiratory deterioration in the form of tachypnea. Subsequently, he was transported to the Emergency Center of our hospital. High-signal lesions were observed from the cervical spinal cord to the thoracic spinal cord on T2-weighted spinal magnetic resonance images. Neuromyelitis optica was suspected, and the patient received steroid pulse therapy and immunoadsorption plasmapheresis. Serum samples obtained upon transfer were positive for anti-aquaporin-4 antibodies, which confirmed the diagnosis of neuromyelitis optica. Thereafter, the patient was transferred to a rehabilitation hospital.

Conclusion: Rapid respiratory failure in neuromyelitis optica is rare, and care is needed while treating these cases.

Key words: Central nervous system, respiration

INTRODUCTION

NEUROMYELITIS OPTICA (NMO) is an inflammatory central nervous system disorder characterized by severe optic neuritis and transverse myelitis. NMO was classified as a separate condition in 2004 following the discovery of an immunoglobulin G antibody specific to it[1] with aquaporin-4 (AQP4), a water channel protein densely expressed in the astrocyte foot process, as its target.[2] The main symptoms of this condition include impaired vision due to optic nerve damage and neuropathy due to spinal cord lesions. Although this disease exhibits a variety of symptoms depending on the localization of nerve lesions in the spinal cord, it rarely involves rapid deterioration of the patient's respiratory conditions to a level where the patient would require a ventilator. Here, we report the case of a patient with NMO who experienced acute respiratory failure.

CASE REPORT

THE PATIENT WAS a 74-year-old man with chronic obstructive pulmonary disease who had a 1-month history of coughing. During this month, he visited a local clinic after developing a fever of 38°C, and was subsequently admitted at another hospital for detailed medical examination. His chest radiograph and computed tomography scan taken upon admission at the other hospital are shown in Figure 1A,B. Despite treatment with antibiotics (sulbactam/ampicillin), he experienced weakness in both lower limbs (manual muscle test [MMT] grade 1–2), pain, and hypoesthesia on day 6 of admission. An emergency magnetic resonance imaging was performed, which revealed high-signal lesions in the center of the spinal cord from C1 to T12 (Fig. 2). Subsequently, the patient was transferred to our hospital for intensive care management due to respiratory deterioration and suspicions of a spinal cord infarction. On examination, we noted weakness in both lower limbs (MMT

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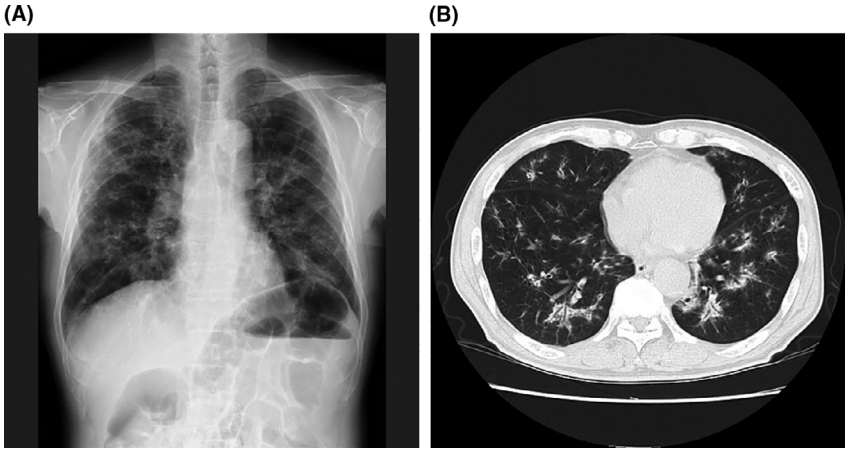

Fig. 1. (A) Chest radiograph taken at another hospital showing infiltrative shadows in both lungs. (B) Chest computed tomography scan taken at another hospital showing scattered mottled nodular shadows in both lungs.

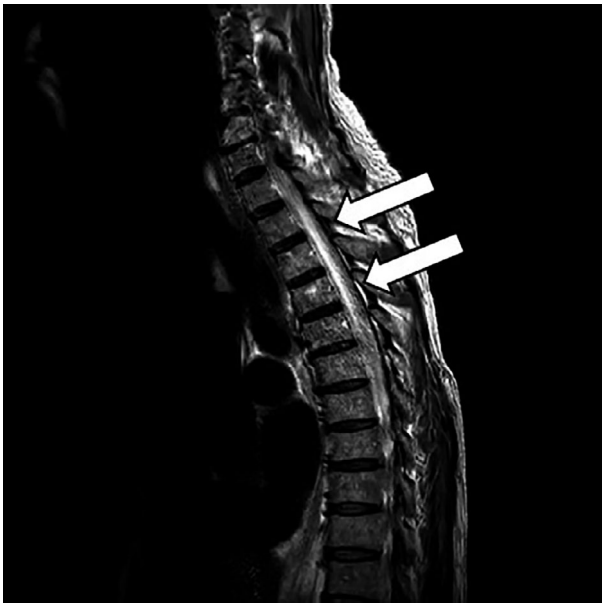

Fig. 2. T2-weighted spinal magnetic resonance image (sagittal) taken at another hospital showing continuous (over 3 or vertebral segments) high-signal areas (arrows) from the cervical spinal cord to the thoracic spinal cord. High signal areas spread from the C7 to Th10 levels with a focus on the medulla.

grade 1 or 2) and pain in the T3 dermatome. In addition, T2-weighted magnetic resonance images taken at our hospital revealed high-signal lesions from the cervical spinal cord to the thoracic spinal cord. NMO was suspected based on the patient's clinical symptoms and the presence of lesions at more than three spinal levels. The patient received steroid pulse therapy between days 2 and 5 of admission, and immunoadsorption plasmapheresis was initiated on day 3 of admission. His respiratory condition gradually deteriorated, and he had CO_2 retention. On day 5 of admission, the patient's respiratory distress worsened despite the initiation of nasal high-flow therapy, and he was intubated. He underwent the second and third rounds of immunoadsorption plasmapheresis on days 7 and 10 of admission, respectively. Thereafter, his respiratory condition gradually improved, and he was extubated on day 13 of admission. The fourth and fifth rounds of immunoadsorption plasmapheresis were performed on days 15 and 18 of admission, respectively. Steroid pulse therapy was administered again from day 16 to day 18 of admission (Fig. 3). The serum sample taken upon transfer to our hospital was positive for anti-AQP4 antibodies, which confirmed the diagnosis of NMO. On day 20 of admission, the patient was transferred to the Department of Neurology at our hospital. Because he was capable of oral feeding without any issues, oral administration of steroids was started to prevent recurrence. Thereafter, his condition stabilized, and he was transferred to a rehabilitation hospital. His MMT grades 4 months after the onset of NMO were 5 in both upper limbs, 2 to 3 in the right leg, and 3 to 4 in the left leg. The patient continues to follow up at the Department of Neurology of our hospital.

DISCUSSION

DEVIC reported the first case of NMO in France in 1984, describing a patient who presented with severe optic neuritis and transverse myelitis.[3] In general, NMO has a female predominance, a tendency to recur, and is prone to co-occur with various autoimmune diseases. NMO tends to be associated with severe optic neuritis and myelitis; it is not

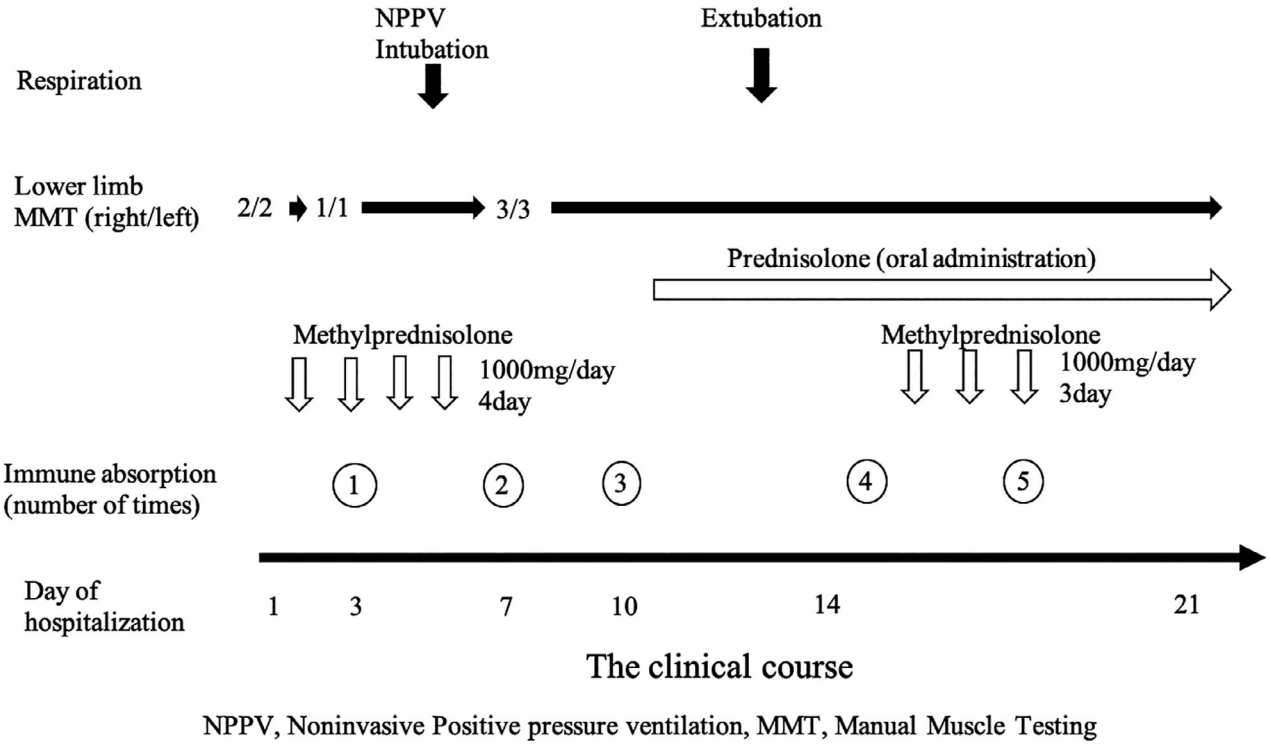

Fig. 3. The clinical course of the case on admission. MMT, manual muscle testing; NPPV, noninvasive positive pressure ventilation.

rare for optic neuritis to lead to blindness, and in many cases, spinal lesions tend to be continuous and span more than three levels. The patients with NMO usually have pain unlike that of spinal infarction. The Wingerchuk criteria published in 2006 are widely used for the diagnosis of NMO.[4] It mentions optic neuritis and myelitis as essential items; however, in 2007, the term "NMO spectrum disorders" began to be used to describe a category of similar diseases, even in patients with optic neuritis or myelitis alone.[5] Cases of NMO, such as this case, with acute respiratory failure are very rare. However, depending on the site of the spinal lesions, there may be cases in which ventilator use is required. In our patient, it was important that we were able to distinguish NMO from spinal cord infarction from the beginning.

Moreover, treatment during the acute exacerbation phase is very important for patients with NMO. Because optic neuritis can cause blindness and myelitis can significantly reduce a patient's ability to perform activities of daily living, treatment must be initiated quickly. The first-line treatment for NMO is steroid pulse therapy. If steroid pulse therapy proves ineffective, at times, clinical improvements can be achieved through immunoadsorption plasmapheresis. Studies have reported that immunoadsorption plasmapheresis may be effective as an acute-phase treatment of NMO.[6] Our patient underwent a total of five rounds of immunoadsorption plasmapheresis (one round ended prematurely), which led to him becoming ventilator independent and eventually being extubated. Furthermore, his ability to perform activities of daily living had improved to the level of wheelchair use by the time he was transferred. Although cases exhibiting prolonged respiratory failure with advancement of nerve lesions are not rare, those with acute respiratory failure are very rare. There are some reports of patients with transient interstitial changes in the lungs due to NMO spectrum disorders.[7] At the time of transfer to our hospital, our patient was receiving antibiotic treatment for pneumonia. NMO was suspected due to the rapid emergence of symptoms in the extremities, but there is a great deal of urgency associated with this condition given the fact that it is not well known in the field of critical care and that it progresses rapidly. Because of the necessity of quickly administering immunoadsorption plasmapheresis, this condition should be considered during the differential diagnosis of patients presenting with respiratory failure and weakness of the extremities without any history of trauma. It is possible that removal

of the anti-AQP4 antibody improved the nerves, which affected the respiratory muscles.

CONCLUSION

HERE, WE REPORTED the case of a patient with NMO who required short-term use of a ventilator after rapid development of respiratory failure.

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DISCLOSURES

Approval of the research protocol: N/A.
Informed consent: We obtained written informed consent from the patient.
Registry and the registration no. of the study/trial: N/A.
Animal study: N/A.
Conflict of interest: None.

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