INTRODUCTION

Neuromyelitis optica spectrum disorders (NMOSD, previously known as neuromyelitis optica or Devic’s syndrome) are rare autoimmune diseases involving the optic nerve, spinal cord, and central nervous system (CNS) [1-3]. Autoantibodies against aquaporin-4 immunoglobulin G (AQP4-IgG) are important in the pathogenesis of NMOSD. The diagnostic criteria were based on the typical clinical, immunological, and radiological features [2]. Brain MRI of NMOSD patients shows typically nonspecific white matter alterations, such as patches and dots, in the subcortical and deep white matter [4]. Brain lesions are often observed as multiple enhanced mass; therefore, differentiating from multiple brain tumors, such as brain metastases, lymphomas, and high-grade gliomas, is necessary. The accurate diagnosis of NMOSD through detailed medical history, laboratory test, and radiologic findings may reduce the rates of false diagnosis and unnecessary test or treatment.

CASE REPORT

We report a rare case of NMOSD, a disease of antibody-mediated disorder, mimicking multiple brain tumors.

A 53-year-old woman, who was diagnosed with multiple brain tumors in a local clinic, was referred to our hospital for further diagnosis and treatment. She had originally visited the clinic for weakness and paresthesia in her right arm and leg. Upon admission, brain MRI showed about 10 multiple brain tumors, which had to be differentiated from multiple brain metastases, lymphoma, and high-grade glioma in both hemispheres. No primary cancer was found in the chest-abdomen-pelvis CT. Subsequent spine MRI revealed multifocal cord signal change involving C2–T7, suggesting myelitis. A decrease in visual acuity was noted when taking a medical history, and optic neuritis was diagnosed upon ophthalmologic examination. With clinical and radiological appearances, multiple brain and spinal cord lesions have been diagnosed as NMOSD. Steroid and immunosuppressive drugs were administered. We should consider the possibility of an autoimmune disease, such as NMOSD, involving the optic nerve, spinal cord, and central nervous system when multiple hemispheric tumefactive lesions are observed.

Key Words Neuromyelitis optica spectrum disorders; Optic neuritis; Spinal cord; Tumor.
from several years ago. Upon ophthalmologic examination, she was diagnosed with optic neuritis. Laboratory examination showed normal findings including negative AQP4-IgG. With clinical characteristics involving the optic nerve, spinal cord, and brain, and typical radiological appearances of the lesions, the multiple brain lesions were diagnosed as NMOSD. Intravenous corticosteroid was administered to treat an acute attack. As maintenance treatment, oral steroid and immunosuppressive drug (azathioprine) have been administered. A 2-year follow-up brain MRI reveals the disappearance of the previously enhanced lesions, but T2 high signal lesions are still observed (Fig. 3).

The Institutional Review Board of Kyungpook National University Hospital exempted informed consent due to its retrospective nature and minimal risk for harm to the patient, and this report was conducted according to the guidelines of the Declaration of Helsinki for biomedical research.

**DISCUSSION**

NMOSD is a rare autoimmune disorder involving the optic nerve, spinal cord, and CNS [1-3]. In the past, it was considered as a form of multiple sclerosis (MS). Now, NMOSD can be distinguished from MS, based on clinical, radiological, and immunological findings. AQP4-IgG test is an important diagnostic tool, but in the novel classification, both groups
(with and without AQP4-IgG) are eligible for NMOSD diagnosis [1,2]. The first international consensus criteria published in 2015 based the diagnosis of NMOSD on the presence of core clinical characteristics, AQP4 antibody status, and MRI features (Table 1) [2]. The diagnostic criteria for NMOSD with negative or unknown AQP4-IgG status are more exacting. This patient was diagnosed through the diagnostic criteria for NMOSD with negative or unknown AQP4-IgG status.

Neurological symptoms, such as encephalopathy and brainstem alteration, develop in 15% of NMOSD patients [5]. Brain lesions are observed in the brain MRI of about 89% of AQP4-negative NMOSD patients [6]. In these patients, the accurate diagnosis through brain MRI findings is very important to differentiate between AQP4-negative NMOSD and other autoimmune diseases and MS [5]. The most common findings in NMOSD were nonspecific small dots and patches in the subcortical and deep white matter. In 9–36% of NMOSD patients, contrasts-enhancing brain lesions are reported [6]. Differentiating these brain lesions from multiple brain tumors is important, as in this case. Characteristic brain MRI findings in NMOSD include periependymal lesions surrounding the ventricles, hemispheric tumefactive lesions, involvement of corticospinal tracts, and cloud-like enhancement [4]. A poorly marginated, subtle, and multiple patchy pattern was called cloud-like enhancement. Brain lesions are generally located close to the ventricles, in the diencephalon and hypothalamus, which are not characterized by central veins. Moreover, cortical lesions are absent in NMOSD [7].

The spine MRI shows characteristic longitudinally extensive lesions (more than three vertebral segments) and central/
gray involvement in the spinal cord of NMOSD patients. Sometimes, continuous lesions in the spinal cord appear as contrast-enhanced lesions in NMOSD patients, which are often misdiagnosed as a malignant spinal cord tumor; thus, a surgical biopsy is performed. Of course, it occasionally occurs in the brain.

Although optic nerve involvement is an important clinical finding for the diagnosis of NMOSD, diagnosing optic neuritis in brain MRI is difficult. Optic neuritis in NMOSD patients may show normal brain MRI, as in this case [8]. In patients with multiple brain lesions mimicking brain tumors, checking whether ophthalmologic symptoms exist is important, even if brain MRI findings show a normal optic nerve. In addition, if abnormal findings on the spine MRI taken after checking the medical history of the spine are found, the NMOSD diagnosis becomes more conclusive.

Discussing the typical radiological findings of NMOSD is of great help in differentiating NMOSD from other diseases, which can reduce unnecessary examinations, such as surgical biopsies. Multiple brain lesions in both hemispheres are likely to be mistaken for malignant brain tumors. To differentiate NMOSD from multiple brain tumors, brain and spine MRI should always be performed. In our study, the entire CNS MRI (brain and spine) was very important to help distinguish them from other diseases. Metastatic brain tumors were a first impression even prior to the spine MRI examination.

As was the case in our patient, NMOSD does not require surgical treatment. The accurate diagnosis of NMOSD was achieved through characteristic brain and spine MRI, ophthalmic examination, and medical history without pathological examination. The precise review of brain and spine MR images in particular can lead to the correct diagnosis of NMOSD.

In conclusion, we report a rare case of NMOSD mimicking multiple brain tumors. NMOSD should be considered in the differential diagnosis of multiple brain lesions in both hemispheres. Sufficient medical history and spine as well as brain MRI are essential in differentiating NMOSD from multiple brain tumors.

Conflicts of Interest
The authors have no potential conflicts of interest.
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None

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