A Case of Anti-aquaporin-4 Antibody-Seronegative NMO Spectrum Disorder with Baló Concentric Lesions

Key words: Baló concentric sclerosis, neuromyelitis optica, aquaporin-4, multiple sclerosis, myelitis, optic neuritis

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The Authors Reply

We thank Dr. Zhou and colleagues for their interest in our article. They suggested that a diagnosis of neuromyelitis optica spectrum disorder (NMOSD) should not be made if anti-aquaporin-4 (AQP4) antibodies are not detected in the serum. Furthermore, they recommended that we diagnose our patient with multiple sclerosis (MS).

First, Wingerchuk et al. defined the NMO spectrum as comprising NMO, limited forms of NMO (single/recurrent longitudinal extensive myelitis and recurrent/bilateral simultaneous optic neuritis), Asian optic-spinal MS, optic neuritis/myelitis associated with brain lesions typical of NMO in their original article on NMOSD (1) cited by Dr. Zhou. Therefore, the article states that anti-AQP4 antibody positivity is not a prerequisite for a diagnosis of NMOSD. In fact, in previous reports, anti-AQP4 serostatuses have been studied in patients with NMOSD (2), and the term “seropositive NMOSD” has been used (3).

We believe that Wingerchuk et al. implied that NMO-related disorders include a very broad spectrum of conditions. Based on their implication, we referred to NMOSD in our article in order to allow readers to comprehend the possible association between NMO and Baló concentric sclerosis.

Second, we disagree that our patient’s diagnosis was MS due to the dissemination of lesions in space and time and the findings of anti-AQP4 antibody negativity. According to the definition of NMO (4), patients with both optic neuritis and myelitis with long extensive spinal cord lesions and brain MRI findings not meeting the MS diagnostic criteria are classified as having NMO, irrespective of the detection of anti-AQP4 antibodies in their serum. The recommended diagnostic process by Dr. Zhou would result in the misdiagnosis of relapsing seronegative NMO cases as MS. Moreover, bilateral simultaneous optic neuritis, which is part of the spectrum of NMOSD, and centrally located myelitis with more than one vertebral segment spinal cord lesion are not typical of MS, as we described in our article.

As Dr. Zhou indicated, the original definition of NMOSD has been associated with the condition of anti-AQP4 antibody seropositivity in order to prevent the inclusion of NMO-unrelated disorders in most previous studies of NMOSD (2). Therefore, we will use the term “partial NMO,” defined as either optic neuritis or transverse myelitis, with positive NMO-IgG (anti-AQP4 antibody) serum, as proposed by Mandler (5) in place of NMOSD in our case-controlled studies in order to prevent any ambiguity in the diagnosis of NMOSD.

The authors state that they have no Conflict of Interest (COI).

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