

Lupus Related Longitudinal Myelitis

To the Editor:

We read with interest the report by Zotos and colleagues describing “longitudinal myelitis” in a patient diagnosed with systemic lupus erythematosus (SLE)¹. The magnetic resonance images demonstrate “longitudinally extensive transverse myelitis” (LETM), which is defined as an inflammatory spinal cord lesion that extends contiguously over 3 or more vertebral segments². This pattern is strongly suggestive of neuromyelitis optica (NMO), a central nervous system (CNS) inflammatory demyelinating disease that causes optic neuritis and myelitis, and that is associated with a highly specific serum autoantibody (NMO-IgG) that targets aquaporin-4^{3,4}. The reported lesion is further suspicious for NMO because it extends into the brain stem, where, as in the reported case, it may cause neurogenic respiratory failure². Finally, the patient’s rapid and significant clinical improvement after corticosteroid and plasmapheresis therapy is also characteristic of NMO.

NMO is often associated with multiple coexisting autoantibodies and clinically evident systemic autoimmunity, including autoimmune thyroid disease, Sjögren syndrome, systemic lupus erythematosus (SLE), myasthenia gravis, and several others⁵. We have shown, using the high specificity of NMO-IgG, that the NMO syndrome (optic neuritis and myelitis) coexists with these systemic autoimmune disorders rather than necessarily being caused by them⁶. Patients with SLE and other systemic autoimmune diseases are uniformly NMO-IgG-seronegative unless they have the clinical NMO syndrome. Unlike antinuclear antibody, extractable nuclear antigen, and other such antibody studies, which are frequently detectable in otherwise healthy individuals or nonspecifically in patients with background autoimmunity, NMO-IgG is extremely specific for NMO. About 70% of patients with clinical NMO are seropositive for NMO-IgG regardless of whether they have detectable autoantibodies, clinically confirmed SLE, or other autoimmune diseases. Therefore, we believe that many cases of “lupus myelitis” are, in fact, NMO with coexisting systemic autoimmunity. The autoantibody and complement profile noted in the case presentation is entirely consistent with NMO.

It therefore is of interest to know the following regarding the case presented by Zotos and colleagues: (1) Which SLE diagnostic criteria were fulfilled? (2) Did she undergo serum testing for NMO-IgG? (3) What did the cerebrospinal fluid analysis reveal (many NMO patients have significant pleiocytosis, sometimes neutrophilic, in the acute myelitis phase)? (4) Was the brain imaging otherwise normal (it usually is in early NMO)? (5) Was the patient being treated for SLE when the LETM occurred? If so, with what immunotherapy?

Accurate and early diagnosis of NMO is critical. Detection of serum NMO-IgG in a patient with first-ever LETM is highly predictive of relapsing CNS disease; more than 50% of patients relapse with myelitis or develop optic neuritis within 1 year of followup⁷. Because of the devastating expression of these attacks, we recommend initiation of immunosuppressive therapy with drugs such as azathioprine, mycophenolate, or rituximab for 5 years after the first such neurological event. We submit that this patient should undergo NMO-IgG testing; if seropositive, she requires longer term immunotherapy and close neurological followup.

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