The patient was treated initially with methylprednisolone followed by 5 cycles of plasma exchange, with minimal improvement in upper extremity muscle strength and sensation despite repeat MRI showing partial resolution of the previously seen lesions. Subsequently she received 4 weekly cycles of IV rituximab therapy. Follow-up at 7 and 14 months demonstrated normal strength in her upper extremities with decreased reflexes, and she was able to walk with use of a walker. Moreover, the patient regained complete right eye vision with improved left eye vision (20/400), clinically consistent with a monophasic process. Repeat NCS/EMG at 7 months revealed improvement, and at 14 months NCS were normal and EMG was significantly improved (table). Repeat AQP4 antibody testing was negative at 7 months posttreatment.

Discussion. Until recently, NMO was believed to be confined to the optic nerves and spinal cord, with no involvement of the peripheral nervous system (PNS). Routine EMG/NCS are not commonly indicated for patients with NMO, so the prevalence of PNS damage coexisting with this disorder is unknown. There have been 5 case reports of seropositive NMO with possible involvement of the PNS,2–4 speculated to result from AQP4 antibody–mediated astrocyte dysfunction or complement activation associated with peripheral AQP4 antibody targets.5 However, to the best of our knowledge, this association has not been described in the setting of seronegative NMO phenotype disease.

Recent studies have shown that some AQP4 antibody–seronegative patients have antibodies against myelin oligodendrocyte glycoprotein (MOG) and other proteins.6 Conceivably, although MOG is localized to the CNS, other targeted complement-activating antibodies might damage the PNS similarly to the mechanism hypothesized for AQP4 antibodies.1,6 Moreover, fulminant NMO spectrum disorders (NMOSDs) are characterized by the presence of severe inflammation, evidenced by neutrophilic pleocytosis and elevated protein in the CSF.7 Reduced local blood flow, hypoxia, and the blood–brain barrier destruction from severe inflammation may partly explain the involvement of the peripheral nerves. Coexistence of multiple autoimmune diseases with NMOSDs has been recently recognized.7 Similar to other immune-mediated diseases,
NMO may be a manifestation of a genetic predisposition to multiple autoimmune conditions. Immune reconstitution in the face of acute viral syndromes, medications, or other causes for immune activation has also been well-described in patients with HIV, as well as in transplant patients following tapering of immunomodulation. It is unknown whether simultaneous onset of the peripheral and CNS involvement in our patient results from an identical antigen targeting 2 different parts of the neuraxis. Lastly, peripheral B cell-mediated antibody activation of the complement system, inflammatory neuronal damage, and necrosis in NMOSDs may benefit from treatment agents like plasmapheresis and rituximab. It is possible that a common mechanism, likely autoimmune in nature, caused the concurrent damage to the central and peripheral neural elements seen in our patient.

This report highlights the association of NMOSDs with peripheral neuropathies and polyradiculopathies. It is essential that clinicians are aware of this unusual association and obtain NCS/EMG whenever there is a clinical suspicion for concomitant involvement of peripheral nerves with NMOSD.

We have included representative nerves that were highly affected by the disease and have shown robust improvement with treatment.

**Table**  Compound motor and sensory nerve action potential amplitudes before and after immunomodulating therapies

<table>
<thead>
<tr>
<th>Site</th>
<th>Normal values</th>
<th>Pretreatment</th>
<th>Posttreatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left ulnar anti sensory (wrist)</td>
<td>&gt;20 µV</td>
<td>13.8 µV</td>
<td>22.5 µV</td>
</tr>
<tr>
<td>Right sural anti sensory</td>
<td>&gt;6 µV</td>
<td>0 µV</td>
<td>4.6 µV</td>
</tr>
<tr>
<td>Left ulnar motor (wrist)</td>
<td>&gt;6 mV</td>
<td>2.6 mV</td>
<td>6.9 mV</td>
</tr>
<tr>
<td>Left median motor (wrist)</td>
<td>&gt;6 mV</td>
<td>3.2 mV</td>
<td>8.1 mV</td>
</tr>
<tr>
<td>Right peroneal motor (ankle)</td>
<td>&gt;2 mV</td>
<td>0.1 mV</td>
<td>2.5 mV</td>
</tr>
<tr>
<td>Right tibial motor (ankle)</td>
<td>&gt;4 mV</td>
<td>0.1 mV</td>
<td>3.6 mV</td>
</tr>
</tbody>
</table>

We have included representative nerves that were highly affected by the disease and have shown robust improvement with treatment.

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Author contributions: Study concept and design: Drs. Feyissa, Shanina, Shah, and Smith. Acquisition of data: Drs. Feyissa, Shanina, Shah, and Smith. Analysis and interpretation of data: Drs. Feyissa, Shanina, and Smith. Drafting of the manuscript: Drs. Feyissa, Shah, and Shanina. Critical revision of the manuscript for important intellectual content: Drs. Feyissa, Shanina, and Smith. Study funding: No targeted funding reported.

Disclosure: A.M. Feyissa, E. Shanina, and R. Shah report no disclosures. R.G. Smith received research support from the University of Texas system. Go to Neurology.org/nn for full disclosure forms. The Article Processing Charge was paid by the authors.

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Received November 19, 2014. Accepted in final form January 20, 2015.

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Neuromyelitis optica phenotype associated with therapy-responsive acute peripheral neuropathy
Anteneh M. Feyissa, Elena Shanina, Rahul Shah, et al.
Neurol Neuroimmunol Neuroinflamm 2015;2;
DOI 10.1212/NXI.0000000000000083

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