A 57-year-old woman presented with a 20-year history of progressive lower extremity weakness, spasticity, and proprioception deficits. She was given a diagnosis of primary progressive multiple sclerosis at age 38. Her Achilles tendons were enlarged (figure 1). Brain MRI was normal. Spine MRI demonstrated T2-hyperintense signal involving the posterior and lateral columns (figure 2). Serum cholestanol level was elevated. CYP27A1 gene sequencing revealed 2 pathogenic variants, c.1183C>T(p.Arg395Cys) and c.410G>A(p.Arg137Gln), confirming the diagnosis of cerebrotendinous xanthomatosis (CTX). Spinal xanthomatosis is a rare variant of CTX presenting with progressive corticospinal and posterior column signs. Early treatment with chenodeoxycholic acid may improve outcomes.

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**Disclosure**
The authors report no disclosures relevant to this manuscript. Go to Neurology.org/N for full disclosures.
Figure 2 Spinal MRI

T2-weighted cervical sagittal (A), thoracic sagittal (B), and axial (C) images demonstrate T2-hyperintense signal abnormality involving the posterior and lateral columns from C2 to T8–T9.

Appendix Authors

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
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</tr>
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<tbody>
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References

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Cristina Valencia-Sanchez, Dean M. Wingerchuk and Radhika Dhamija
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