A 58-year-old man presented with a 3-year history of minimally progressive right-sided weakness. Examination revealed facial asymmetry, mild pyramidal weakness in the upper and lower limbs with spasticity, brisk reflexes, and upgoing plantar on the right only. Sensory

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Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.
examination was normal. MRI brain abnormalities are demonstrated (figures 1 and 2 and video). Serial EMG of the limbs and motor unit number estimation were symmetrically normal without evidence of concurrent lower motoneuron (LMN) pathology. Unilateral midbrain atrophy in Mills syndrome is rarely reported.1 The current case illustrates the associated asymmetric reduction in corticospinal tract fibers and absence of LMN involvement, further supporting a hemiparetic variant of primary lateral sclerosis.2

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**Disclosure**
The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

**Appendix**

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<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
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**References**

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