A 63-year-old woman had slowly progressive dementia. EEG displayed periodic sharp wave complexes. MRI showed cortical ribbon pattern in cortex and hyperintensity in basal ganglia (figure, A–H). Genetics testing confirmed homozygosity for methionine at the polymorphic codon 129 of the PRNP gene. At 34 months after disease onset, she was alive and had not reached akinetic mutism. Skin prions were positive by real-time quaking-induced conversion for detection of prion: positive in 4 duplicate wells.

Figure Sequential diffusion-weighted imaging changes

Transverse scans through the lower part of paracentral lobe (A–D) and the basal ganglia (E–H) at different periods after disease onset are shown. Hyperintensities were initially constrained to the posterior parietal lobe, and extended to the anterior parietal lobe. (I) Real-time quaking-induced conversion for detection of prion: positive in 4 duplicate wells.

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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.
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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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