

Long survival sporadic Creutzfeldt-Jakob disease

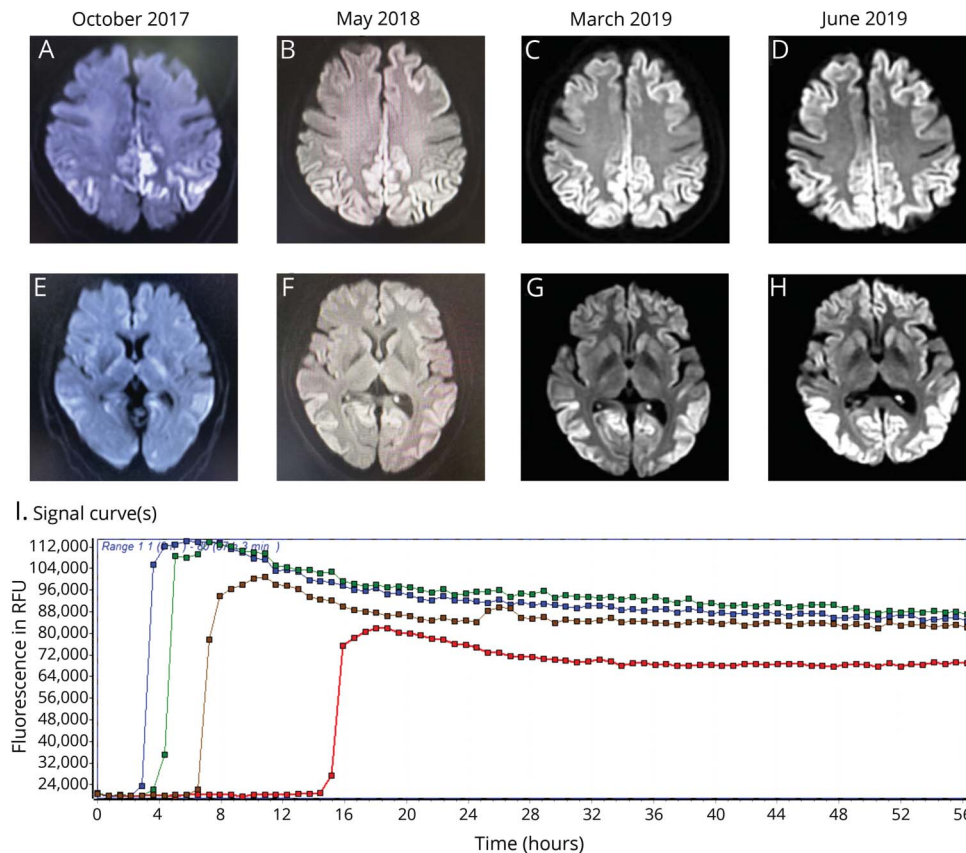
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Figure Sequential diffusion-weighted imaging changes



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.

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 assays (figure, I). This case demonstrates a rare long-survival sporadic Creutzfeldt-Jakob disease case that is contradictory to traditional duration of illness and the utility of minimally invasive skin biopsy testing for antemortem confirmation.^{1,2}

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Disclosure

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

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Name	Location	Contribution
Xiao-Ying Liu, MD, PhD	Department of Neurology, Ruijin Hospital, Shanghai Jiaotong University School of Medicine, Shanghai, China	Drafting/revising the manuscript, analysis or interpretation of data, acquisition of data

Appendix (continued)

Name	Location	Contribution
Qi Shi, PhD	National Institute for Viral Disease Control and Prevention, Chinese Center for Disease Control and Prevention, Beijing, China	Analysis or interpretation of data, acquisition of data
Gang Wang, MD, PhD	Department of Neurology, Ruijin Hospital, Shanghai Jiaotong University School of Medicine, Shanghai, China	Drafting/revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data, study supervision

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