Teaching *NeuroImages*: Extensive cortical involvement in Creutzfeldt-Jakob disease

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Figure 1 MRI brain

(A–F) Low-grade restricted diffusion (white arrows) affecting the cerebral cortex. (G, H) Mild cerebral atrophy seen on fluid-attenuated inversion recovery images.

A 53-year-old alcoholic man presented with a 2-day history of worsening confusion. Initial examination showed appendicular hypertonia with multifocal arrhythmic asynchronous myoclonic jerks suggestive of cortical myoclonus occurring spontaneously intermittently and stimulus sensitive, with exaggerated deep tendon reflexes and extensor plantars. MRI brain demonstrated low-grade restricted diffusion affecting the entire cerebral cortex (figure 1), sparing the subcortical gray matter and cerebellum.¹ T1-weighted imaging and fluid-attenuated inversion recovery sequences showed mild cerebral atrophy. 14-3-3 protein testing was positive and generalized slowing was demonstrated on EEG.² Changes consistent with Creutzfeldt-Jakob disease (CJD) were found on histopathology (figure 2). The patient had no risk factors for familial, new variant, or iatrogenic CJD. Sporadic CJD was the final diagnosis.

AUTHOR CONTRIBUTIONS

Sanjay Hettige: study concept and design, acquisition of data, analysis of data, interpretation of data, drafting of manuscript. Monica Badve: study concept and design, interpretation of data, study supervision, revision of manuscript for intellectual content. Manisha Narasimhan: study concept and design, interpretation of data. Lynette Masters: acquisition of data, analysis of data, interpretation of data. Shu Wang: acquisition of data, analysis of data, interpretation of data.

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DISCLOSURE

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Figure 2 Microscopic histopathology

(A) Frontal cortex with spongiform change consistent with Creutzfeldt-Jakob disease. The underlying white matter (WM) is unaffected (hematoxylin & eosin, $\times 20$). (B) Frontal cortex with positive immunohistochemical staining for anti-prion antibody (clone 12F10, $\times 20$). GM = gray matter; S = sulcus.

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