Teaching *NeuroImages*: Hemispheric-onset Creutzfeldt-Jakob disease with concordant MRI and EEG findings

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Figure 1 MRI at presentation

The axial diffusion-weighted image (A) showed restricted diffusion throughout the cortical ribbon of the right hemisphere. There was corresponding decreased signal on the apparent diffusion coefficient map. The changes were not easily seen on the fluid-attenuated inversion recovery sequence (B).

A 73-year-old woman presented with 6 weeks of progressive confusion and visual hallucinations. On examination, she had moderately impaired attention,

concentration, and memory; mild left hemiparesis; dysmetria; and visual and sensory neglect. No myoclonus was observed. MRI showed restricted diffusion along the right hemispheric cortical ribbon (figure 1). Periodic sharp waves were lateralized to the right hemisphere on EEG (figure 2A). Protein 14–3–3 was detected in the CSF. Creutzfeldt-Jakob disease (CJD) was confirmed by brain biopsy (figure 2B).

Although CJD is typically associated with diffuse cerebral pathology,¹ it can present with unilateral neurologic symptoms and concordant MRI and EEG findings.

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Figure 2 EEG and findings on brain biopsy

EEG (A) demonstrated right hemispheric background attenuation and 1-Hz periodic sharp waves (characteristic findings in Creutzfeldt-Jakob disease [CJD]). Left hemisphere background was relatively preserved. Brain biopsy (B) showed diffuse spongiform changes and coalescent vacuoles (inset, white arrows). Immunostain and Western blot (PrPSc) confirmed prion disease, most consistent with CJD.

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