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, it can present with unilateral neurologic symptoms and concordant MRI and EEG findings.

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REFERENCE

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