Teaching NeuroImages: DWI and EEG findings in Creutzfeldt-Jakob disease

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A 62-year-old woman, previously healthy, presented with 3 weeks of progressive short-term memory loss, dyscalculia, dysgraphia, appetite loss, and frequent episodic left arm dystonic posturing and eyebrow elevation. On examination, she also had finger agnosia, dysarthria, agraphesthesis, and astereognosis bilaterally, with myoclonus, ideomotor apraxia (left worse), and wide-based gait. EEG showed slowing and periodic lateralized discharges over the right hemisphere with triphasic morphology, less often involving the left, reflecting clinical asymmetry (figure 1). MRI brain showed diffusion restriction in the bilateral basal ganglia with cortical ribboning more prominent on the right involving several gyri, consistent with Creutzfeldt-Jakob disease (figure 2).

Author contributions
Dr. Ganesh: concept, acquisition of data, and writing the initial drafts of the manuscript. Dr. Hoyte: concept, acquisition of data, and writing the initial drafts of the manuscript. Dr. Agha-Khani: acquisition of data and critical revision of manuscript for intellectual content. Dr. Yeung: concept and critical revision of manuscript for intellectual content.

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Disclosure
A. Ganesh serves on the Neurology® Resident & Fellow Section’s editorial board. L. Hoyte, Y. Agha-Khani, and M. Yeung report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

References
Figure 1 EEG findings of asymmetric slowing and periodic lateralized discharges in Creutzfeldt-Jakob disease

Representative epochs of the EEG show (A) posterior alpha rhythm on the left vs theta-delta slowing over the right hemisphere and (B) periodic lateralized discharges over the right hemisphere with triphasic morphology.

Figure 2 MRI findings of cortical and striatal diffusion restriction in Creutzfeldt-Jakob disease

Axial sections on (A) diffusion-weighted imaging, (B) apparent diffusion coefficient, (C) T2-weighted, and (D) T1-weighted MRI sequences show true diffusion restriction in the bilateral caudate and putamen and multiple gyri, more on the right (cortical ribboning), but without other changes on T2 or T1.
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