Focal photoparoxysmal response in the Heidenhain variant of CJD

Hidden from view!

Figure EEG and radiologic findings in the Heidenhain variant of Creutzfeldt-Jakob disease

Axial diffusion-weighted MRI showing hyperintensity (arrow) in the left occipital cortex (A). Fluorodeoxyglucose-PET demonstrating marked hypometabolism in the bilateral occipital lobes, maximal on the left (B). EEG during sleep depicting periodic left parietooccipital spikes (arrows) (C) and left occipital photoparoxysmal response (arrows) during low-frequency stimulation (D).

A 73-year-old, right-handed man presented with new-onset right visual field diminution and visual hallucinations. Examination showed a right homonymous hemianopia without cognitive impairment. Brain imaging showed left occipital cortex abnormalities (figure, A and B). EEG showed sleep-predominant, periodic left occipital spikes and an occipital photoparoxysmal response (figure, C and D). A second-generation RT-QuIC (real-time quaking-induced conversion) test detected pathogenic prion proteins in the CSF, confirming Creutzfeldt-Jakob disease (CJD).

The Heidenhain variant of CJD¹ is characterized by visual disturbances as the presenting feature and marked neuropathologic abnormalities involving the occipital cortices. EEG findings include occipital and generalized periodic sharp complexes.² Sleep activation and photoparoxysmal activity have not been reported.

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Study funding: No targeted funding reported.

Disclosure: The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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Neurology 2016;86;1647-1648

DOI 10.1212/WNL.0000000000002617

This information is current as of April 25, 2016

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