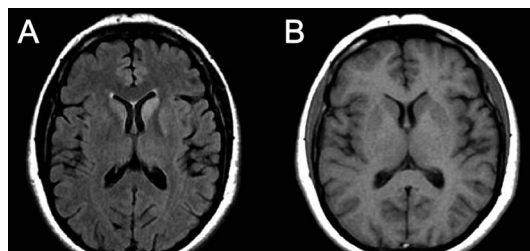


Teaching NeuroImages: Pseudohypertrophic cerebral cortex in end-stage Creutzfeldt-Jakob disease

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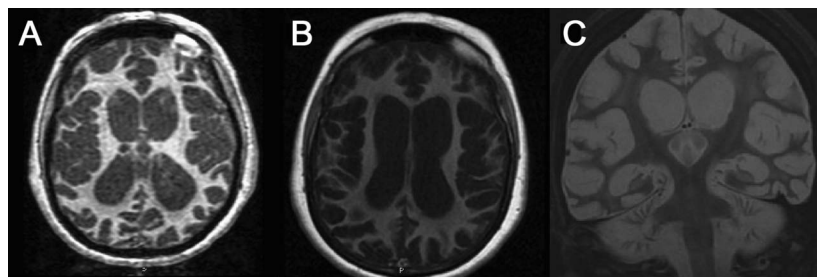
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Figure 1 MRI at admission



Axial fluid-attenuated inversion recovery image (A) showing signal hyperintensity in the head of caudate nuclei, in left lentiform nucleus, and in medial frontal lobe cortex bilaterally. Axial T1-weighted image (B) was normal.

Figure 2 MRI 1 year later



Axial fluid-attenuated inversion recovery (A), T1-weighted (B), and coronal T2-weighted (C) images revealing marked gyral atrophy and thickening of cerebral cortex with CSF-like signal and enlargement of lateral ventricles (pseudohypertrophic cortex). Diffuse white matter hyperintensity is also evident.

A 43-year-old woman presented with 1 month of progressive lower limb burning sensation, blurred vision, and gait disturbance. Her mother died of Creutzfeldt-

Jakob disease (CJD). Neurologic examination revealed only cerebellar ataxia. EEG revealed periodic generalized discharges. MRI (figure 1) showed fluid-attenuated inversion recovery (FLAIR) hyperintensity in caudate and lenticular nuclei. Molecular analysis confirmed genetic CJD (*PRNP* E200K mutation). One month later, she became comatose. After 10 months, EEG showed diffuse background flattening without periodic abnormalities, and MRI (figure 2) disclosed diffuse pseudohypertrophy of cerebral cortex. MRI in this patient with end-stage CJD allowed us to reveal in vivo the macroscopic spongiform changes usually observed at autopsy.^{1,2}

AUTHOR CONTRIBUTIONS

S. Gasparini and E. Ferlazzo: drafting/revising the manuscript for content, including medical writing for content; study concept or design. D. Branca, A. Labate, V. Cianci, and M.A. Latella: analysis or interpretation of data. U. Aguglia: drafting/revising the manuscript for content, including medical writing for content; study concept or design; study supervision or coordination.

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DISCLOSURE

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

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