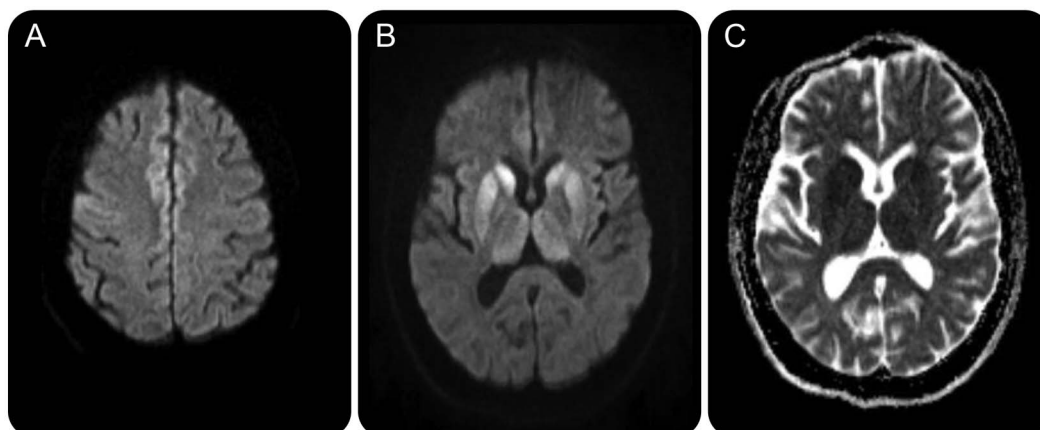


Supranuclear gaze palsy and horizontal ocular oscillations in Creutzfeldt-Jakob disease



Figure MRI diagnostic of Creutzfeldt-Jakob disease



MRI brain diffusion-weighted imaging with hyperintense cortical gyri (A), basal ganglia and pulvinar diffusion restriction (B), with apparent diffusion coefficient dropout (C).

A 59-year-old patient presented with marked cognitive impairment. Examination revealed supranuclear gaze palsy and ocular oscillations (video at Neurology.org). Extremity and axial muscle tone was increased, accompanied by full body tremulousness suggestive of polymyoclonus. Motor abnormalities had progressed over 3 months but cognitive problems developed in 1–2 weeks. MRI (figure) and CSF (positive real-time quaking-induced conversion) were diagnostic of Creutzfeldt-Jacob disease (CJD).

Supranuclear ophthalmoplegia is a known feature of CJD but when combined with parkinsonism can lead to a misdiagnosis of progressive supranuclear palsy.¹ Pendular oscillations, not previously described in CJD, are typically due to failure of brainstem eye movement neural integrators.²

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Supplemental data
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