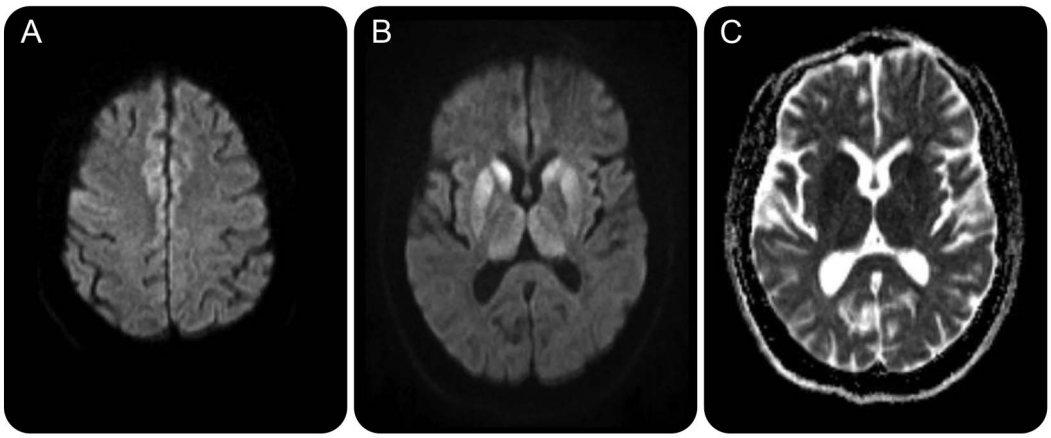
Supranuclear gaze palsy and horizontal ocular oscillations in Creutzfeldt-Jakob disease

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]

Asya I. Wallach, MD, Hannah Park, MD, Janet C. Rucker, MD, Horacio Kaufmann, MD

From NYU School of Medicine and Bellevue Hospital, New York.

Supplemental data at Neurology.org

Author contributions: Asya Wallach: clinical care of the patient, study concept, acquisition of data, writing the manuscript. Hannah Park: clinical care of the patient, acquisition of data. Janet Rucker: acquisition of data, analysis and interpretation of data, study supervision, critical revision of the manuscript for intellectual content. Horacio Kaufmann: study concept and supervision, critical revision of the manuscript for intellectual content.

Study funding: Supported in part by funding from the NIH (U54NS065736 and 1U01NS078025-01).

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

Correspondence to Dr. Wallach: Asya.Wallach@nyumc.org

1. Petrovic IN, Martin-Bastida A, Massey L, et al. MM2 subtype of sporadic Creutzfeldt-Jakob disease may underlie the clinical presentation of progressive supranuclear palsy. J Neurol 2013;260:1031–1036.
2. Gresty MA, Ell JJ, Findley LJ. Acquired pendular nystagmus: its characteristics, localising value and pathophysiology. J Neurol Neurosurg Psychiatry 1982;45:431–439.

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

Asya I. Wallach, Hannah Park, Janet C. Rucker, et al.

Neurology 2017;89;749
DOI 10.1212/WNL.0000000000004242