Progressive ataxia and palatal tremor

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A previously healthy 72-year-old man presented with 1 year of progressive ataxia and slurred speech. There was no notable family history and no history of alcohol abuse or neurotropic drug use. Examination showed bilateral hypometric saccades, a 2–3 Hz palatal tremor with face and neck involvement, and truncal ataxia (video at Neurology.org/cp). There was no pendular nystagmus. MRI brain 6 months after the onset of symptoms showed high signal abnormality in the inferior olivary nuclei of the anterior medulla and mild superior vermal atrophy (figure, A and B). His clinical presentation and neuroimaging were consistent with a diagnosis of progressive ataxia and palatal tremor (PAPT), a rare idiopathic neurodegenerative disease with bulbar features, palatal tremor, and cerebellar ataxia. PAPT imaging features include a typical hypertrophic olivary appearance on MRI, sometimes with cerebellar degeneration.^{1,2}

In most cases of oculopalatal tremor, the lesion originates in the central tegmental tract, which connects the red nucleus and deep cerebellar nuclei projections to the inferior olives. Olivary hypertrophy occurs as secondary phenomenon from deafferentation, with MRI demonstrating T2-weighted hyperintensity and initial hypertrophy followed by atrophy typically several months later. However, in sporadic PAPT, the exact location of the lesion in

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Figure T2-weighted MRI

(A) Axial T2-weighted image demonstrates bilateral high signal in the area of the inferior olivary nuclei. (B) Sagittal T2-weighted image demonstrates hyperintensity of the inferior olivary nuclei.

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uncertain. In patients who present with progressive ataxia or palatal tremor, neuroimaging is indicated and can help to identify this uncommon diagnosis.

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AUTHOR CONTRIBUTIONS

Clotilde Hainline: drafting/revising the manuscript. Andreas Neophytides: critical revision of the manuscript, selecting and collecting video. Maria J. Borja: critical revision of the manuscript, selecting and collecting images. Steven L. Galetta: critical revision of the manuscript.

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