

Teaching Video NeuroImages: A Triad of Tremor, Ataxia, and Cognitive Impairment

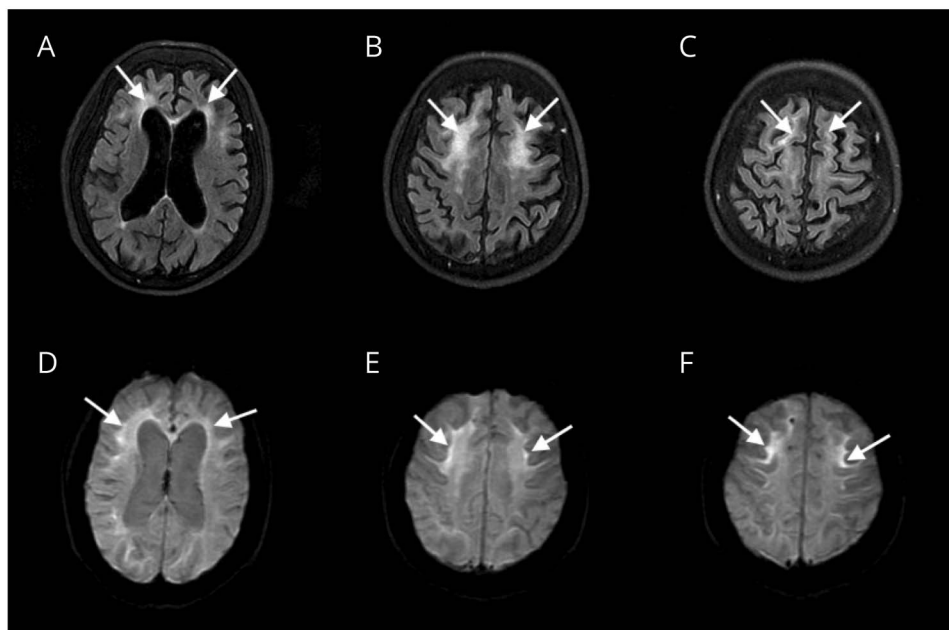
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Figure 1 MRI Brain



Axial MRI brain performed at age 72 years showed bilateral subcortical white matter lesions (arrows) on axial fluid-attenuated inversion recovery images (A–C) and high signal intensity along the corticomedullary junction (arrows) on diffusion-weighted images (D–F).

A 67-year-old woman presented with a 2-year history of forgetfulness and unsteadiness. She scored 17 on the Montreal Cognitive Assessment. She had tremor of head and upper limbs (video 1) since early adulthood, which was diagnosed as essential tremor. Brain MRI showed leukoencephalopathy and high signal intensity along the corticomedullary junction on diffusion-weighted images (figure 1). The diagnosis of adult-onset neuronal intranuclear inclusion disease (NIID) was confirmed by skin biopsy showing eosinophilic intranuclear inclusions (figure 2).¹ NIID is a clinically heterogeneous rare neurodegenerative disease.² Its characteristic MRI pattern should prompt confirmation of the diagnosis by skin biopsy.

Study Funding

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

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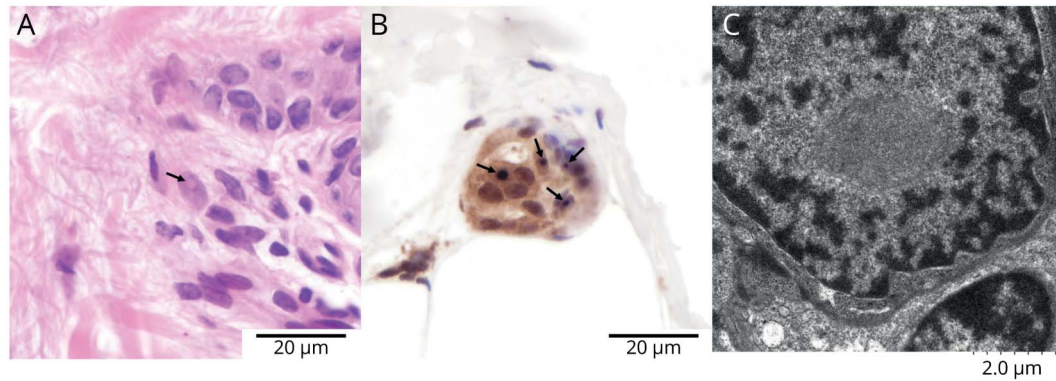
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Figure 2 Skin Biopsy



Hematoxylin & eosin stain of fibroblasts (A) and ubiquitin immunostain of sweat gland cells (B) showing intranuclear inclusions (arrows); the inclusions consisted of electron-dense filamentous materials without membrane structure on electron microscopy (C).

Appendix Authors

Name	Location	Contribution
Lisa W.C. Au, FHKAM	The Chinese University of Hong Kong	Designed and conceptualized study, analyzed the data, drafted the manuscript for intellectual content
Ko Ho, PhD	The Chinese University of Hong Kong	Interpreted the data
Joshua J.X. Li, MBChB	The Chinese University of Hong Kong	Interpreted the data

Appendix (continued)

Name	Location	Contribution
Vincent C.T. Mok, MD	The Chinese University of Hong Kong	Interpreted the data, revised the manuscript for intellectual content

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