Teaching Video Neuroimages: A triad of tremor, ataxia and cognitive impairment

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A 67-year-old female presented with two year history of forgetfulness and unsteadiness. She scored 17 on the Montreal Cognitive Assessment (MoCA). She had tremor of head and upper limbs (video, http://links.lww.com/WNL/B294) 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 (DWI) (figure 1). The diagnosis of adult-onset neuronal intranuclear inclusion disease (NIID) was confirmed by skin biopsy showing eosinophilic intranuclear inclusions (figure 2).\textsuperscript{1} NIID is a clinically heterogeneous rare neurodegenerative disease.\textsuperscript{2} Its characteristic MRI pattern should prompt confirmation of the diagnosis by skin biopsy.

Appendix 1: Authors

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Lisa W.C. Au, FHKAM</td>
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<td>Design and conceptualized study; analyzed the data; drafted the manuscript for intellectual content</td>
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Teaching Slides - http://links.lww.com/WNL/B293
Video - http://links.lww.com/WNL/B294

References:
Figure 1 legend

MRI brain. Axial MRI brain performed at age 72 years showed bilateral subcortical white matter lesions (arrows) on axial fluid-attenuated inversion recovery (FLAIR) images (A-C), and high signal intensity along the corticomedullary junction (arrows) on diffusion-weighted (DWI) images (D-E).
Figure 2 legend

**Skin biopsy.** H&E 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).

Video legend

Video recorded at age 72 years. The patient showed tremor of head and upper limbs that were present at rest and worsened on posture and movement. She failed tandem walking.
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