Teaching Video NeuroImages: Myoclonus as the presenting feature of Wilson disease

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A 10-year-old boy with no pertinent medical or family history developed repeated falls and progressive gait and speech decline over 1 year. Examination revealed multifocal myoclonus and generalized dystonia (video). EEG lacked epileptiform activity. Kayser-Fleischer rings, serum ceruloplasmin of 6 mg/dL (normal 20–60 mg/dL), and 24-hour urinary copper of 108.94 μg (normal 15–60 μg) confirmed Wilson disease (WD). MRI brain revealed T2 and fluid-attenuated inversion recovery hyperintensity in basal ganglia, thalami, brainstem, and right frontal cortex, with the latter showing diffusion restriction (figure). The patient improved.

Figure Basal ganglia, thalamus, brainstem, and cortical involvement on MRI of the brain

MRI of the brain shows fluid-attenuated inversion recovery hyperintensity in basal ganglia (A), thalami (A), midbrain (B), dorsal pons (C), and right frontal cortex (D, white arrow). Diffusion-weighted imaging (E, white arrow) and apparent diffusion coefficient maps (F, white arrow) show a small area of diffusion restriction in the right frontal cortex.
neurologically with zinc and penicillamine therapy. Myoclonus is uncommon in WD, with multifocal myoclonus at onset rarely reported.2

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N. Kumar: conception, design, and writing the first manuscript. D. Kumar: review and critique.

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