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

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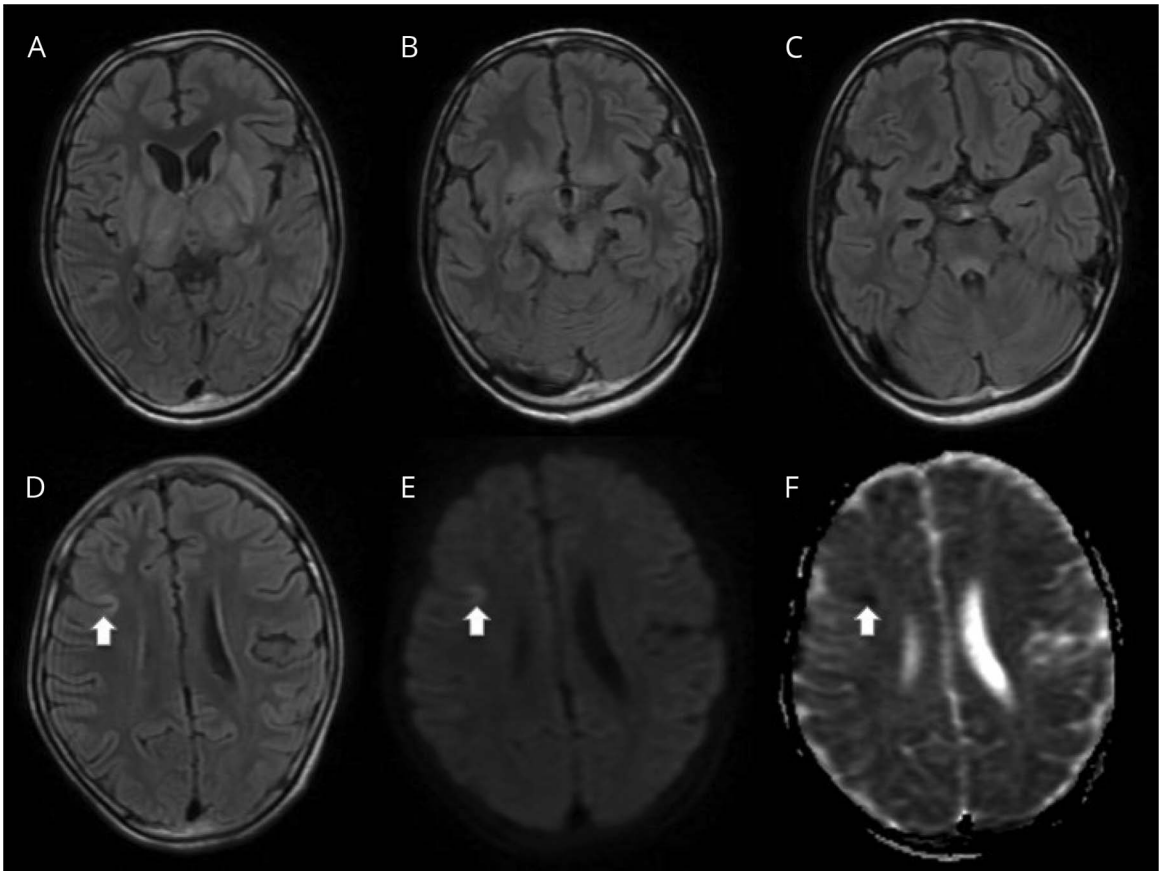
Figure Basal ganglia, thalamus, brainstem, and cortical involvement on MRI of the brain



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.

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

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neurologically with zinc and penicillamine therapy. Myoclonus is uncommon in WD,[1] with multifocal myoclonus at onset rarely reported.[2]

Author contributions

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

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Disclosure

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