Teaching Video NeuroImages: Excessive grinning in Wilson disease

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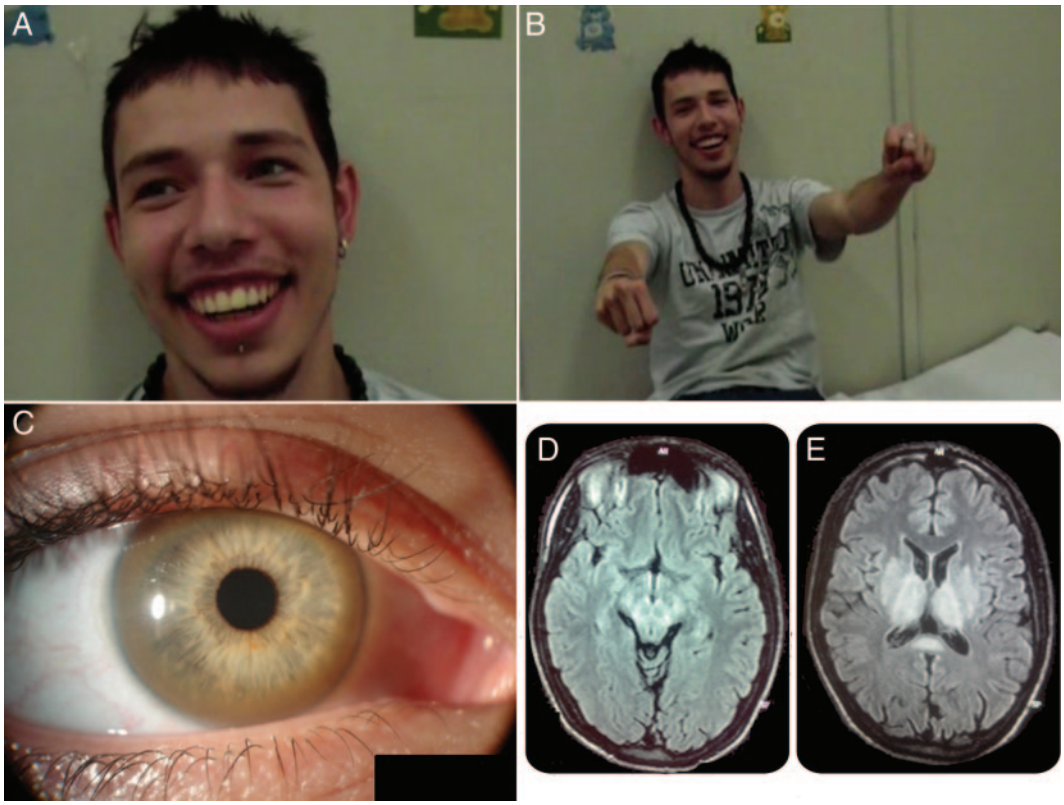
Figure (A, B) Excessive grinning, (C) Kayser-Fleischer ring, (D) "face of giant panda" sign and basal ganglia T2 hyperintensity



A 19-year-old man presented with a 3-month history of excessive grinning. Examination revealed unrestrained grinning and mild symmetric parkinsonism. Wilson disease was suspected and confirmed by the presence of Kayser-Fleischer ring (figure), suggestive brain MRI (figure), low ceruloplasmin, and high urinary copper levels.

Wilson disease is a disorder of copper metabolism characterized by hepatic impairment and movement disorders. Typical facial manifestations, although not pathognomonic, include excessive grinning, in which the patient grins to trivial stimuli,[1] as demonstrated in this report; sustained open-mouth smile, when a parkinsonian face is associated with a dystonic dropped jaw (sometimes referred to as "vacuous smile")[2]; and fixed forced smile, when facial dystonia produces a sustained spasm of risorius and zygomaticus muscles (also referred to as "risus sardonicus").

REFERENCES

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2. Das SK, Ray K. Wilson's disease: an update. Nat Clin Pract Neurol 2006;2:482–493.

Supplemental data at www.neurology.org

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