Teaching Video NeuroImages: Hand tremor, tongue and perioral fasciculation in a patient with Kennedy disease

A 54-year-old man presented with a 10-year history of progressive gait difficulty, muscle weakness, and fatigue. Clinical examinations revealed mild nasal speech; postural hand tremor; absence of deep tendon reflexes; atrophy and fasciculation in tongue, facial (perioral), and limb muscles; and gynecomastia (video on the Neurology® Web site at Neurology.org). Nerve conduction studies had normal results; however, needle EMG demonstrated a neurogenic pattern with spontaneous activity (fibrillation, positive sharp wave, and fasciculation) in cranial and limb muscles. Genetic testing confirmed (CAG expansion of the androgen receptor gene on X chromosome) the diagnosis of spinobulbar muscular atrophy or Kennedy disease, an X-linked adult-onset degenerative disorder of motor neurons.1,2

AUTHOR CONTRIBUTIONS
Dr. Mohammad Rohani: acquisition of data, drafting and revising the manuscript. Dr. Shahnaz Miri: drafting and revising the manuscript.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
Teaching Video NeuroImages: Hand tremor, tongue and perioral fasciculation in a patient with Kennedy disease
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Neurology 2015;84:e76
DOI 10.1212/WNL.0000000000001340

This information is current as of March 9, 2015

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