Teaching Video NeuroImages: Palatal myoclonus in leukodystrophies
A clinical sign orienting to Alexander disease

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A 22-year-old man with Alexander disease type II diagnosed by a compatible MRI with bilateral white matter hyperintensities and brainstem atrophy (figure) and mutation c.236G>A (p.Arg79His) in the GFAP gene presented with recent onset continuous palatal myoclonus without ear clicking (video 1).

Palatal myoclonus is caused by a lesion in the triangle of Guillain-Mollaret (formed by dentate nucleus, red nucleus, and inferior olivary nucleus) and associated with hypertrophic olivary degeneration.1 As Alexander disease is a leukodystrophy that predominantly affects the brainstem, palatal myoclonus can be a useful sign to distinguish it from other leukodystrophies.2

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

Figure Brain MRI

(A) Sagittal T1 MRI brain shows atrophy of brainstem (arrow) and cerebellum (asterisk). (B) Axial fluid-attenuated inversion recovery sequence shows extensive white matter hyperintensities.

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### References


### Appendix

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