Teaching Video NeuroImages: Palatal myoclonus in leukodystrophies

A clinical sign orienting to Alexander disease

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Neurology® 2020;94:e230-e231. doi:10.1212/WNL.0000000000008795

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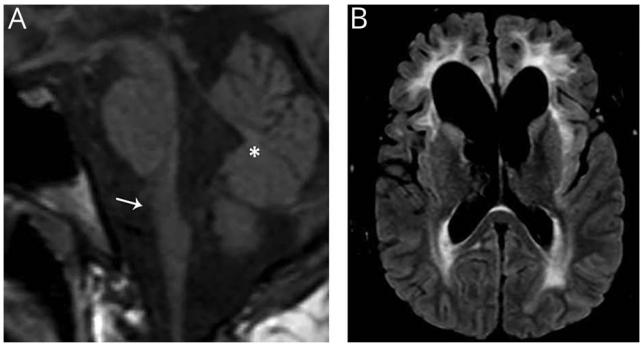
Figure Brain MRI



(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.

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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Video

Study funding
No targeted funding reported.

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

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Name	Location	Role	Contribution
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References

1. Pearce JMS. Palatal myoclonus (syn palatal tremor). Eur Neurol 2008;60:312–315.
2. Howard KL, Hall DA, Moon M, Agarwal P, Newman E, Brenner M. Adult-onset Alexander disease with progressive ataxia and palatal tremor. Mov Disord 2008;23:118–122.

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