

Teaching Video NeuroImages: Palatal myoclonus in leukodystrophies

A clinical sign orienting to Alexander disease

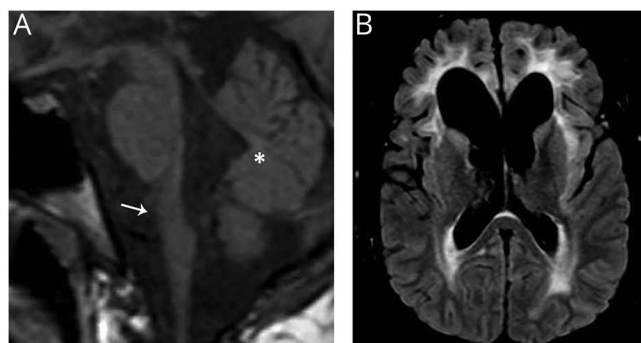
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Figure Brain MRI



(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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Disclosure

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▶ Video

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Name	Location	Role	Contribution
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José Bernardo Escribano-Paredes, MD	Neurology Department, Ramón y Cajal University Hospital, Madrid, Spain	Author	Revised the manuscript for intellectual content
Sebastián García-Madrona, MD	Neurology Department, Ramón y Cajal University Hospital, Madrid, Spain	Author	Revised the manuscript for intellectual content

Appendix *(continued)*

Name	Location	Role	Contribution
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Adriano Jiménez-Escrig, MD, PhD	Neurology Department, Ramón y Cajal University Hospital, Madrid, Spain	Author	Major role in the acquisition of data, revised the manuscript for intellectual content

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