Teaching Video NeuroImages: Anti-IgLON5 Disease

A Long-Course Presentation With a Response to Immunotherapy

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A 74-year-old woman presented with a 5-year history of progressive gait instability with recurrent falls, dysphonia, dysphagia, and sleep problems, which included insomnia, breathing difficulties, and complex vocalizations during sleep (talking, singing, laughing). Examination showed bilateral ptosis, upward gaze palsy, hoarseness, dysprosody, and gait instability (video 1). No parkinsonism, chorea, or dementia was detected.

Brain MRI and PET as well as CSF measures were unremarkable. Video-polysomnography revealed a disorganized sleep architecture but not parasomnia. Anti-IgLON5 antibodies were positive in CSF and serum by cell-based assay of HEK cells transfected with IgLON5 and immunohistochemistry on rat brain. Human leukocyte antigen genotyping detected HLA-DQB1*05:01.

Immunotherapy with IV methylprednisolone 1 g/d for 5 days and immunoglobulins resulted in improvement of gait and speech over a month (video 1). A second course of immunoglobulins provided clinical stability.

Anti-IgLON5 disease represents a paradigm of autoimmune neurodegeneration with core features of a specific sleep disorder, bulbar symptoms, and gait abnormalities.¹ Its recognition is important because some cases respond to immunotherapy.²

Study Funding

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

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Name	Location	Contribution
Javier Villaceros-Álvarez, MD	Hospital Universitario Rey Juan Carlos; Hospital Infanta Elena; Hospital General de Villalba, Madrid, Spain	Designed and conceptualized the study, major role in acquisition of data, drafted the manuscript for the intellectual content, literature review
Carlos Manuel Ordás, MD	Hospital Universitario Rey Juan Carlos; Hospital Infanta Elena; Hospital General de Villalba, Madrid, Spain	Major role in acquisition of data, critical review of the manuscript for intellectual content
Gustavo Torres-Gaona, MD	Hospital Universitario Rey Juan Carlos; Hospital Infanta Elena; Hospital General de Villalba, Madrid, Spain	Major role in acquisition of data, critical review of the manuscript for intellectual content
Ana Díez-Barrio, MD	Hospital Universitario Rey Juan Carlos; Hospital Infanta Elena; Hospital General de Villalba, Madrid, Spain	Major role in acquisition of data, critical review of the manuscript for intellectual content

Appendix (continued)

Name	Location	Contribution
Cristina Prieto-Jurczynska, MD	Hospital Universitario Rey Juan Carlos; Hospital Infanta Elena; Hospital General de Villalba, Madrid, Spain	Major role in acquisition of data, critical review of the manuscript for intellectual content
Carles Gaig, PhD	Hospital Clínic, Barcelona, Spain	Major role in acquisition of data, critical review of the manuscript for intellectual content

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