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

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### References


### Appendix: Authors

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### Appendix (continued)

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