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**Anti-IgLON5 Disease: A Long-course Presentation with a Response to Immunotherapy**

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Supplemental: Consent form signed by the patient Consent form signed by the patient's daughter (assistant in the video).

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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, etc). Examination showed bilateral ptosis, upward gaze palsy, hoarseness, dysprosody and gait instability (video1). No parkinsonism, chorea or dementia were detected.

Brain MRI and PET as well as CSF parameters were unremarkable. A 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. HLA-genotyping detected HLA-DQB1\*05:01.

Immunotherapy with intravenous methylprednisolone 1gr/day for five days and immunoglobulins resulted in improvement of gait and speech during a month (video1). 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<sup>1</sup>. Its recognition is important since some cases respond to immunotherapy<sup>2</sup>.

**Video-<http://links.lww.com/WNL/B300>**

**Teaching Slides-<http://links.lww.com/WNL/B301>**

**References:**

1. Gaig C, Graus F, Compta Y, et al. Clinical manifestations of the anti-IgLON5 disease. *Neurology*. 2017;88(18):1736-1743.
2. Cabezudo-García P, Mena-Vázquez N, Estivill Torrús G, Serrano-Castro P. Response to immunotherapy in anti-IgLON5 disease: A systematic review. *Acta Neurol Scand*. 2020;141(4):263-270.

**Video title:**

**Video 1.** Patient affected with anti-IgLON5 disease, pre- and post-immunotherapy.

**Video legend:**

**Video 1.** The patient is describing her symptoms. Note the hoarseness and the dysprosody of the speech, as well as the bilateral ptosis. The oculomotor exam reveals an upward gaze palsy. Gait is very unsteady, requiring an assistance. After immunotherapy an improvement of the speech and gait is detected.

## Appendix 1: Authors

Name	Location	Contribution
Javier Villaceros Álvarez, MD	Hospital Universitario Rey Juan Carlos, Madrid. Spain. Hospital Infanta Elena, Madrid. Spain. 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 Bandera, MD	Hospital Universitario Rey Juan Carlos, Madrid. Spain. Hospital Infanta Elena, Madrid. Spain. Hospital General de Villalba, Madrid. Spain.	Major role in acquisition of data. Critical review of the manuscript for intellectual content.
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Carles Gaig, MD, PhD	Hospital Clínic, Barcelona. Spain.	Major role in acquisition of data. Critical review of the manuscript for intellectual content.

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