Movement Disorders
CLINICAL PRACTICE

CLINICAL VIGNETTE

Unusual Head Movements in Anti-IgLON5 Disease

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Anti-IgLON5 disease is a clinically heterogenous condition characterized by prominent sleep disorders, preceded or accompanied by neurological signs and symptoms, and the presence of IgLON5 antibodies.[1] We present a case of a fluctuating segmental dystonia and sleep-related movement disorder as the main presentation of anti-IgLON5 disease.

A 65-year-old woman presented to our outpatient clinic complaining of an involuntary forward-flexed position of her head that had progressed during the past 3 months. She described an uncomfortable sense of traction in her throat and chest that seemed to pull her head anteriorly. No sensory trick was reported. A new-onset orthopnoea forced her to sleep upright. Upon further questioning, she mentioned insomnia and fatigue, fluctuating dysarthria and dysphagia, falls, hypotension, and mild short-term memory problems for about a year.

Upon inspection, a marked antecollis independent of body position or activity was present, although a normal head position could still be achieved upon command and when supine. A laterocollis was infrequently present, probably induced by voluntary muscle contraction (looking to the camera) (Video S1). While walking, a dystonic lateroflexion of the trunk emerged (Video S2), closely resembling Parkinson's disease–related Pisa syndrome. Apart from the presence of mildly slurred speech and subtle hypomimia, physical and neurological examination was unremarkable and more important, no axial muscle weakness was detected.

Polysomnography revealed severe obstructive sleep apnoea, fragmented sleep consisting only of stage 2 and 3 non-rapid eye movement sleep and a reduced total sleep time. During relaxed wakefulness in transition to sleep, semirhythmic head flexion-extension movements were noticed. In sleep stages 2 and 3, these movements occurred in clusters with a maximum duration of 223 seconds, usually lasting around 30 seconds (Video S3). Background electroencephalogram, brain magnetic resonance imaging and routine cerebral spinal fluid analyses were normal. This combination of neuropsychiatric symptoms, movement disorders, and sleep abnormalities prompted us to test for IgLON5 antibodies in addition to our center's testing panel for immune-mediated encephalitis. They were positive in a serum sample (not tested on cerebral spinal fluid), and HLA-DRB1*10:01 and HLA-DQB1*05:01 genotypes, known to be associated with this disease, were present.

Although several treatments (corticosteroids, plasmapheresis, rituximab) were initiated, none proved beneficial. On the contrary, her symptoms progressively worsened.

Although cervical dystonia has been described in case series,[2] our patient illustrates that unusual head movements may be the presenting feature of anti-IgLON5 disease. Although no sensory trick was present, we believe that the movements can be attributed to segmental dystonia affecting cervical and truncal musculature.

The exact nature of the remarkable sleep-related movements remains unclear. There is a certain resemblance with headbanging, a sleep-related rhythmic movement disorder, usually seen in children and consisting of repetitive movements involving neck and trunk muscles emerging predominantly during wake–sleep transitions, that rarely persist during sleep.[3] Because the sleep-related movements in our patient were semirhythmic and largely occurred during stages 2 and 3, we suggest describing them as "atypical headbanging."[4]

Our case illustrates that a diagnosis of anti-IgLON5 disease should be considered in patients presenting with segmental dystonia, especially when accompanied by sleep-related movement disorders.■

Author Roles

(1) Clinical Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

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Disclosures

Ethical Compliance Statement: An informed consent was obtained with written confirmation from the patient that she gave her permission to submit this work and use the video, in which the patient can be identified. The consent stipulates that authorization has been obtained in compliance with any laws regarding patient authorizations relating to the use or disclosure of protected health information of the jurisdiction(s) to which the patient and the physician are subject. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines. No IRB approval was necessary.

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Supporting Information

Supporting information may be found in the online version of this article.

Video S1 Antecollis without clear sensory trick. The presence of laterocollis can be debated because it is probably attributed to voluntary contraction looking at the camera.

Video S2 Lateroflexion of the trunk observed while walking.

Video S3 Fragment of polysomnographic video during sleep stage 3, showing atypical headbanging.