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LETTER TO THE EDITOR

Orthostatic Myoclonus as a Presentation of Hashimoto Encephalopathy

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Dear Editor,

Orthostatic myoclonus (OM), which was recently described, is a clinical presentation characterized by abundant muscle jerks in the lower extremities that appear immediately upon standing. It can also present as unsteadiness during gaiting or standing, which leads to sudden falls and gait initiation difficulty or gait apraxia.[1] OM can appear in various diseases, such as atypical parkinsonism, subcortical microvascular encephalopathy, radiculopathy, tumor and autoimmune disease.[2] Here, we report a case of Hashimoto encephalopathy (HE) presenting as OM.

A 56-year-old female was referred to the Department of Neurology in Haeundae Paik Hospital complaining of tremulousness in both legs upon standing. She had a history of hypertension, spinal stenosis, and hyperthyroidism and was taking antihypertensive medication and methimazole.

About a year ago, she experienced paroxysmal shaking of both hands, which lasted approximately 5 minutes each and occurred 5–6 times a day. The symptoms improved naturally without any treatment. However, one year later, she complained of her legs shaking when standing. The symptoms started abruptly 15 days ago.

On examination, myoclonic jerks were clearly noted in both legs immediately upon standing. She showed unsteadiness on standing and had difficulty initiating gaiting. Myoclonic jerks were evident in both hands and legs. There were no other neurologic deficits (Supplementary Video 1 in the online-only Data Supplement).

Because she had a history of hyperthyroidism, laboratory tests, including thyroid hormone levels and antithyroid antibodies, were performed. Routine blood counts, renal function tests, liver function tests, and glucose were within normal limits. Her free thyroxine level was elevated (2.46 ng/dL), while her thyroid stimulating hormone (TSH) was decreased (0.01 mIU/L). All of the antithyroid antibodies were significantly increased. Her TSH receptor antibody was over 40.0 IU/L (normal range: ≤ 1.5 IU/L), thyroid stimulating antibody was 565.9% (normal range: < 140.0%), anti-thyroglobulin antibody (anti-Tg Ab) was 959.9 IU/mL (normal range: 0–4.1 IU/mL), and anti-thyroid peroxidase antibody (anti-TPO Ab) was over 1,000 IU/mL (normal range: 0–5.6 IU/mL). Other laboratory tests were unremarkable, and test results for other serum autoimmune antibodies were negative.

Brain magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) studies were performed. There was no hydrocephalus or other neurodegenerative changes on MRI. The CSF was also clear with normal levels of red blood cells, white blood cells, protein, and glucose.

Additionally, surface electromyography (sEMG) was recorded from the rectus femoris of the patient to confirm OM. Irregular, intermittent myoclonic bursts appeared when the patient was standing with a brief median duration of 50 ms (Supplementary Video 2 in the online-only Data Supplement).

We postulated that the OM was a neurological manifestation of HE. She was administered 750 mg of intravenous methylprednisolone for 3 days with oral tapering for 6 days. She showed a considerable response to steroid therapy and was then trans-

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ferred to the Department of Rehabilitation Medicine in Haeundae Paik Hospital for rehabilitation. One month later, at the follow-up outpatient appointment, she showed a normal gait, and the follow-up sEMG showed no signs of any remaining OM. There was no relapse, and she is currently leading a normal life (Supplementary Video 3 in the online-only Data Supplement).

OM was first described in 2007 by Glass et al.,[1] who reported 15 patients who presented with myoclonic jerks exclusively during an upright posture. OM is difficult to discriminate from orthostatic tremor (OT), which also shows oscillatory movement in the lower extremities when standing. Electrophysiologic studies, including sEMG, in OM show nonrhythmic, synchronous, short duration (less than 100 ms) bursts recorded from the lower limb muscles during standing. However, sEMGs of OT patients show rhythmic, asynchronous, high frequency 16–18 Hz discharges.[3] Our patient presented with trembling of both legs upon standing. Since the movement was difficult to distinguish with only our naked eyes, we first concluded that the patient presented with OT. However, the sEMG findings of this patient showed nonrhythmic bursts with a short duration (50 ms), which fulfilled the proposed criteria for OM.[1] Our case also showed mild myoclonic movement of both upper extremities, simultaneously with OM. In a review of the clinical phenotype of OM, Glass et al.[1] reported OM with myoclonus of the arms and hands, which is similar to our case. This report supports the hypothesis that OM should be considered a syndrome, not a specific disease.

The underlying etiology of OM remains unclear, but it has been known to be frequently associated with neurodegenerative diseases such as Parkinson's disease, atypical parkinsonism, dementia and systemic disease.[1] Additionally, autoimmune etiologies have previously been proposed in a few patients with OM. A patient with OT superimposed with OM who had an excellent response to intravenous immunoglobulin (IVIG) was reported to have an antibody against an unknown antigen.[4] Another case of OM was proven to be associated with antibodies against contactin-associated protein-2 (Caspr2) with near complete improvement after IVIG.

We first excluded neurodegenerative and structural diseases. Additionally, inflammatory central nerve system disorders were excluded based on the normal CSF findings. The detection of autoimmune antithyroid antibodies and a near complete response to steroids strongly supported the diagnosis of HE in this patient.

HE, otherwise known as steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT), is a rare neurological complication of autoimmune thyroiditis. HE is characterized by encephalopathy with various clinical presentations and elevations in antithyroid antibodies (anti-Tg Ab and/or anti-TPO Ab) in the absence of alternative causes, such as brain tumor, stroke, or infection of the central nervous system. Additionally, the patient's response to steroid therapy is essential for the diagnosis of HE. Although the main clinical presentation of HE is encephalopathy, pure movement disorders without encephalopathy as a manifestation of HE have rarely been reported, such as chorea, dystonia and cerebellar ataxia.[5] We report OM as another clinical manifestation of HE.

Recently, one case of HE presenting with OM has been reported.[6] However, there was no electrophysiological study confirming OM, differentiated from OT. In our case, the movement of the patient mimicked the clinical presentation of OT and was difficult to discriminate without the help of an electrophysiological study. We suggest that OM should be added to the spectrum of symptoms associated with HE and that OM must be confirmed through electrophysiological studies, such as sEMG.

Ethics Statement

Informed consent was secured from the patient for using her video and information for publication purpose.

Supplementary Video Legends

Video 1. This video shows orthostatic myoclonus of the patient upon admission. Myoclonic movement of both legs can be seen when the patient is standing, and unsteadiness of the gait is noted.

Video 2. This video confirms orthostatic myoclonus through surface electromyography (sEMG). sEMG was recorded from the rectus femoris of the patient. Irregular, intermittent myoclonic bursts appeared only when the patient was standing, with a brief median duration of 50 ms.

Video 3. This video was filmed two months after steroid therapy. There are no more myoclonic movements upon standing, and the patient shows a normal gait.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.22146.

Conflicts of Interest

The authors have no financial conflicts of interest.

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Author Contributions

Conceptualization: Jinyoung Youn. Data Curation: Jeong Ik Eun. Formal analysis: Kyong Jin Shin. Methodology: Jongmok Ha. Writing—Original draft: Hyunyoung Hwang. Writing—review & editing: Jinse Park.

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