

Orthostatic Myoclonic Jerks in a Case of Hashimoto's Encephalopathy

Amit Shankar Singh¹  Arshdeep Singh Sidhu² 

¹Department of Neurology, Fortis Hospital, Mohali, Punjab, India

²Department of Internal Medicine, Fortis Hospital, Mohali, Punjab, India

Address for correspondence Amit Shankar Singh, MD, DM, Department of Neurology, Fortis Hospital, OPD Block, Room No. 05, Mohali, Punjab 160062, India (e-mail: amitkgmu@gmail.com).

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Abstract

Keywords

- ▶ myoclonus
- ▶ Hashimoto's encephalopathy
- ▶ antithyroid peroxidase antibodies
- ▶ extrapyramidal
- ▶ jerks

Hashimoto's encephalopathy (HE) is an uncommon syndrome with the characteristic triad of positive antithyroid antibodies (most commonly antibodies to thyroid peroxidase), response to steroids, and clinical picture presenting either as stroke-like pattern of focal neurological deficit or slowly progressive cognitive impairment. Myoclonus or tremors, seizures, and psychosis are other associated features which can be seen in HE. Herein, we report a girl with an uncommon presentation of orthostatic axial and myoclonic jerks in bilateral lower limbs in a case of HE.

Introduction

Hashimoto's encephalopathy (HE) is a rare neurological disorder with a varied clinical spectrum. Commonly, patients present with stroke-like pattern of recurrent, acute to subacute episodes of focal neurological deficit or diffuse, slowly progressive cognitive dysfunction which can manifest as dementia, hallucinations, confusion, or somnolence.¹ Overlap of different types of movement disorders are described in relation with HE. We report a girl, who was unable to walk due to jerky lower body and lower limb movements for 2 years, resulting in her bed-ridden state.

Case Presentation

A 16-year-old girl presented to the clinic with complaints of inability to walk for almost 2 years due to jerky movements in both lower limbs while standing and severe imbalance while walking (see ▶ **Video Segments 1 and 2**). These abnormal jerky movements were aggravated by putting load over her limbs and completely subsiding while sleeping or at rest. Her symptoms

were subacute in onset and progressed over few weeks, hampering her movements completely. Before our assessment, patient was treated as psychosis and conversion disorder, but there was no improvement in her condition.

Video 1

Segment 1 (on first OPD visit) showing bilateral lower limb and axial myoclonic jerks causing postural imbalance. Segment 2 (on admission) showing inability of the patient to stand from sitting position and inability to walk independently due to lower limb myoclonic jerks, which were completely absent in sitting posture. Segment 3 (after 4 weeks of treatment) showing complete absence of lower limb jerks, patient is able to walk properly without any support and even can do tandem walking. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0042-1748176>.

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On examination, she had bilateral lower limb and axial myoclonic jerks which increased on standing, suggesting orthostatic nature of these jerks. These myoclonic jerks led to severe postural imbalance so much so, that she had to take support of others to walk (see ► **Video Segment 2**). No motor weakness or sensory deficits were present. Reflexes were normal. Mild lower limb cogwheel rigidity was present on examination. Rest of physical examination was unremarkable.

Her routine blood investigations were normal. Magnetic resonance imaging of the brain and spinal cord was normal. Electroencephalography and nerve conduction studies were normal. Cerebrospinal fluid (CSF) examination showed mild increase in proteins, and rest was normal. Antinuclear antibodies were negative. Serum copper and ceruloplasmin levels were normal. Autoimmune encephalitis panel in CSF and serum were negative. However, she had T3 of 54 ng/dL (80–200) and T4 of 4.1 µg/dL (5.10–14.10) levels with elevated thyroid-stimulating hormone 16.4 µIU/mL (0.27–4.20). Antithyroid peroxidase (anti-TPO) antibodies were strongly positive (1,200 IU/mL [0–8]).

With this clinical profile, positive anti-TPO antibodies, normal metabolic, infectious, structural and other autoimmune parameters, HE was diagnosed and pulse intravenous steroid therapy (1 g intravenous pulse methylprednisolone once daily for 5 days) was started with levothyroxine supplementation, followed later by oral steroids (1 mg/kg/d for 2 weeks) in tapering doses.

Following 5 days' course of intravenous steroids, patient started responding slowly after third dose, with gradual and steady improvement. Complete clinical resolution was attained with 3 weeks of therapy, which later persisted on oral steroids. Finally, the patient was able to walk without any support with a total treatment duration of 3 to 4 weeks (see ► **Video Segment 3**).

Discussion

HE is a distinctive neurological disorder with specific clinical features in conjunction with high antithyroid antibodies. However, antithyroid antibodies can be seen in the normal population also. Recent articles by Mattozzi et al² and Delgado-García and Balint³ discussed about the controversies surrounding HE. With recent advancements and new revelations in immunology and autoimmune neurological disorders, in future, this entity may be amalgamated with other disorders, but for the time being, any clinical syndrome with very high anti-TPO antibodies, characteristic clinical features, and good response to steroids is regarded as HE.

Since the description by Brain et al,⁴ many new symptoms have been added to the spectrum of HE. Extrapyramidal system involvement leading to varied movement disorders is now being recognized in HE, including myoclonus, tremors, ataxia, dystonia, chorea, tics, palatal tremors, paroxysmal kinesigenic dyskinesia, myorhythmia, and parkinsonism.^{5–7} The index patient had myoclonic jerks, but they were predominantly present in lower limbs and were noted only in

standing posture, suggestive of orthostatic nature of these myoclonic jerks.

Orthostatic myoclonus term was first introduced by Glass et al⁸ in 2007 from Mayo Clinic as a cause of gait impairment in neurodegenerative disorders. Since then, several etiologies have been reported for orthostatic myoclonus, such as Parkinson's disease and atypical parkinsonism, dementia with Lewy bodies, Alzheimer's disease, mild cognitive impairment, normal pressure hydrocephalus, cerebral amyloid angiopathy, mild small vessel cerebral disease, postradiotherapy of resected frontal brain tumor, osmotic demyelination following rapid sodium correction, Lance-Adams' syndrome, and contactin-associated protein-2 positive autoimmune encephalitis.

The exact pathophysiology of orthostatic myoclonus and why jerks only occur while standing in these varied disorders is still not known, but the most plausible reason is the presence of a subcortical generator. A possibility of pontocerebellothalamocortical network involvement is also likely but further investigations such as functional imaging studies in such patients may clearly elucidate the exact pathophysiology of this disorder.

Cortical and subcortical myoclonus have been previously described in HE, but orthostatic myoclonic jerks have not been hitherto reported. Because of these orthostatic myoclonic jerks, the index patient had postural imbalance and she was not able to stand or walk without support. These abnormal movements cannot be categorized as orthostatic tremors as they persisted on walking. The current case illustrates the fact that orthostatic myoclonic jerks can be a presenting rare movement disorder associated with HE and can be treated with standard steroid treatment.

Conflict of Interest

None declared.

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