
Case Report

Orthostatic Myoclonus Presented as Unsteadiness and Gait Disturbance - A Case Report

Meng-Tsang Hsieh, Yu-Ting Lan, Shin-Pin Hsu, Yi-Cheng Tai

Orthostatic myoclonus is a hyperkinetic movement disorder involving the lower limbs upon standing. Gait disturbance and unsteadiness are the most common presentations. Here we described the first case of orthostatic myoclonus in Taiwan. A 67-year-old man presented with subjective unsteadiness and falling. Neurological examination showed irregular but fine muscle jerks over bilateral tibialis anterior muscles. Surface electromyography revealed myoclonic bursts in bilateral lower limbs. Orthostatic myoclonus is a rare but important differential diagnosis of gait disturbance, especially when there is a mismatch between the subjective and objective findings.

Key words: orthostatic, myoclonus, EMG, unsteadiness, fall

Introduction

Myoclonus is a hyperkinetic movement disorder with sudden muscle contraction or lapse.¹ Orthostatic myoclonus is characterized by abundant muscle jerks in multiple leg muscles that appear or increase immediately upon standing.^{2,3} Bursts on electromyography (EMG) characteristic of orthostatic myoclonus present during standing are typically alleviated during sitting or walking.³

We describe an uncommon case of orthostatic myoclonus in a patient with subjective

unsteadiness and gait disturbance.

Case report

A 67-year-old man with hypertension, diabetes mellitus, hyperlipidemia, and chronic kidney disease had unsteadiness and tilting feeling for one week when walking but not sitting, resulting in his falling several times. He did not have dizziness, vertigo, blurred vision, diplopia, limbs numbness, weakness, muscle twitching, or leg jerking. There was also no precipitating factor.

On examination, the visual, motor,

From the ¹Department of Neurology, E-Da Hospital, I-Shou University, Kaohsiung, Taiwan.

Received: February 22, 2016

Accepted: June 3, 2016

Address reprint request and correspondence to: Yi-Cheng Tai, Department of Neurology, E-Da Hospital/I-Shou University, No. 1, Yida Road, Jiaosu Village, Yanchao District, Kaohsiung City 82445, Taiwan.

Tel: +886-7-6150011 ext. 252142, Fax: +886-7-6150940, E-mail: b88401074@ntu.edu.tw

sensory, and coordination systems were all normal. The muscle strength was full in all limbs. He could stand without assistance. However, upon standing, there were irregular and very fine muscle jerks over both lower limbs which disappeared when he was sitting. There was no helicopter sign on auscultation of leg muscles and the posture reflex was intact.

The surface EMG (Fig. 1 and 2) showed short duration (30-50 ms) and arrhythmic bursts (ranging from 2 to 5 Hz) over bilateral tibialis anterior and gastrocnemius muscles upon standing. Similar finding was not

recorded during sitting or with bilateral legs outstretched. His brain CT was normal. No more falling was noted after treatment with oral clonazepam.

Discussion

Orthostatic myoclonus is a symptom rather than a disease. The characteristic abundant lightening-like muscle jerks which appear or increase immediately upon standing is suggestive of the diagnosis. Surface EMG is a useful diagnostic tool because it can detect the brief and irregular orthostatic EMG bursts² with typical duration ranging from 20 to 100 ms and a number of bursts per second^{4,5} characteristic of orthostatic myoclonus.

Orthostatic myoclonus typically presents with involuntary leg movements upon standing. Prolonged standing does not improve but exaggerates the instability. Patients experience complete resolution of their hyperkinesia promptly by leaning forward onto an object or taking weight off of their legs. However, they do not have myoclonus of upper limbs when leaning forward.⁶ The degree of subjective instability is not predictive of orthostatic movement disorder. Patients may not necessarily complain of tremor or shaking limbs when standing still.^{2,4} Orthostatic myoclonus can also manifest itself as gait disturbance.² It is easy to misdiagnose this unsteadiness as pyramidal, extrapyramidal, sensory, or cerebellar system lesion.

Orthostatic myoclonus can be idiopathic or secondary to a systemic or a neurodegenerative disorder.² In one study, 7 out of 15 patients with orthostatic myoclonus had concomitant Parkinson disease, Alzheimer disease, dementia with Lewy body, multiple system atrophy, or amyloid angiopathy² and 2 were accompanied by a systemic illness (i.e., systemic necrotizing vasculitis, chronic renal failure). In another study,⁶ 1 out of 16 was diagnosed with hepatic encephalopathy and the rest were

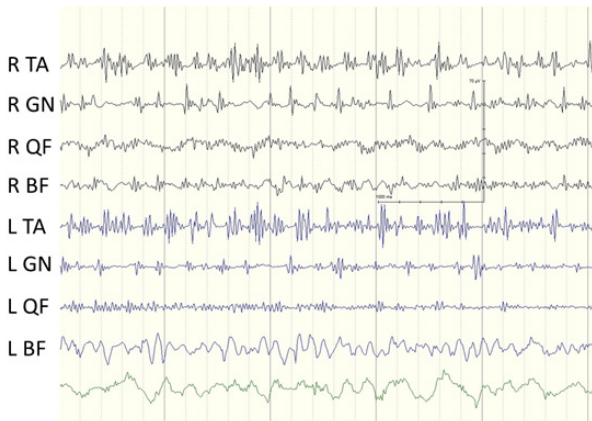


Fig. 1 The surface electromyography (EMG) recorded from lower limbs. When patient was standing showed brief, irregular, multifocal muscle bursts (duration 30-100ms). The frequency of EMG bursting upon standing ranged from 2 to 5 Hz.

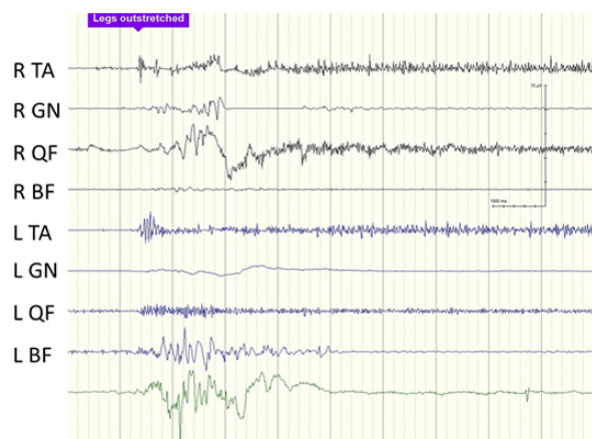


Fig. 2 The surface EMG activity recorded from the same location with the leg outstretched showed no pathological muscle bursts. BF = biceps femoris muscle, GN = gastrocnemius muscle, L = left, QF = quadriceps femoris muscle, R = right, TA = tibialis anterior muscle

associated with neurodegenerative diseases. However, so far there was no long-term data showing the latency to the diagnosis of neurodegenerative disease.

Primary orthostatic tremor is similar to orthostatic myoclonus. Both consist of muscle contractions causing discomfort or unsteadiness during standing and are noted in patients over 65 years of age.⁶ Primary orthostatic tremor presents with intolerable unsteadiness upon standing but patients rarely fall,⁷ while patients with orthostatic myoclonus can stand without assistance but have frequent falls. Orthostatic myoclonus is easily distinguished from primary orthostatic tremor with EMG by the non-rhythmic discharges and the absence of the high frequency discharge (16 to 18 Hz).² The clinical and electrophysiological similarities between these two conditions raise the possibility that abnormal hyperkinetic physiology may progress from tremor to myoclonus, depending upon the natural history of the underlying disease.⁴

Clonazepam is the preferred treatment.² Some antiepileptic drugs, such as levetiracetam or gabapentin, may also help.^{2,6} Non-pharmacological strategy such as gait-assistive device may reduce hyperkinesia when patients lean forward and take some weight off their legs.⁶ However, there is no data on long-term treatment outcome.

Although we did not perform the EMG recording with the patient leaning forward onto an object, the diagnosis was confirmed

by the absence of EMG discharges with legs outstretched. Both conditions involve isometric leg muscle contractions without weight bearing.

In conclusion, our case suggests that detailed history taking, neurological examination, and surface EMG recording are helpful in making differential diagnosis for patients with gait disturbance of undetermined origin. Orthostatic myoclonus may be underdiagnosed but once diagnosed, it is suggested that the patient should be followed for the possible underlying neurodegenerative diseases.

References

1. Espay AJ, Chen R: Myoclonus. *Continuum* 2013; 19:1264-86.
2. Glass GA, Ahlskog JE, Matsumoto JY: Orthostatic myoclonus: a contributor to gait decline in selected elderly. *Neurology* 2007;68:1826-30.
3. Gerschlager W, Brown P: Myoclonus. *Curr Opin Neurol* 2009;22:414-8.
4. Leu-Semenescu S, Roze E, Vidailhet M, et al: Myoclonus or tremor in orthostatism: an under-recognized cause of unsteadiness in Parkinson's disease. *Mov Disord* 2007;22:2063-9.
5. Gasca-Salas C, Arcocha J, Artieda J, et al: Orthostatic myoclonus: an underrecognized cause of unsteadiness? *Parkinsonism Relat Disord* 2013;19:1013-7.
6. van Gerpen JA: A retrospective study of the clinical and electrophysiological characteristics of 32 patients with orthostatic myoclonus. *Parkinsonism Relat Disord* 2014;20:889-93.
7. Sharott A, Marsden J, Brown P: Primary orthostatic tremor is an exaggeration of a physiological response to instability. *Mov Disord* 2003;18:195-9.