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Letter to the Editor

Different orthostatic hyperkinesias go "Thump"

We describe the surface electromyogram (SEMG) and auscultatory findings of a 65-year-old man with probable multiple system atrophy, who presented with gait instability and falls. He was ultimately diagnosed with orthostatic myoclonus (OM) based on SEMG results. OM is an orthostatic hyperkinesia that can induce "shaky legs" and exacerbate gait instability (Glass et al., 2007; Leu-Semenescu et al., 2007). In contrast to previous reports (Brown, 1995; Yaltho and Ondo, 2014) this case demonstrates that auscultation of leg muscles with a stethoscope is likely of little diagnostic value with regard to OT, as it fails to distinguish OT from OM and other orthostatic hyperkinesias.

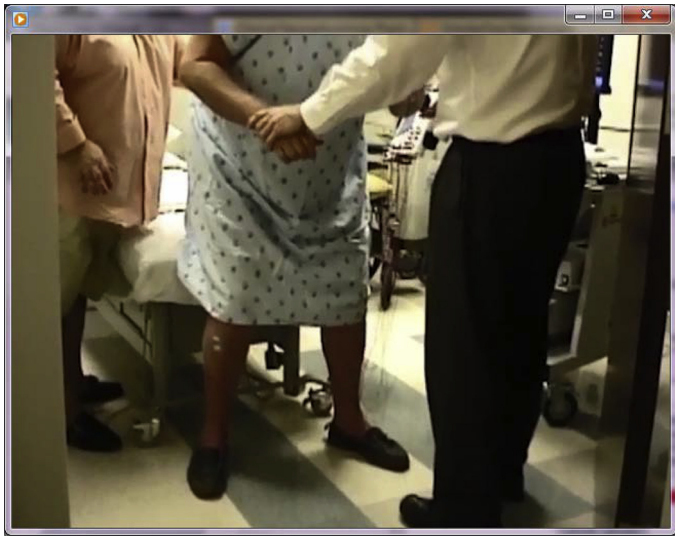
We evaluated a 65-year-old man who presented with progressive gait instability and falls over three years. He was diagnosed with parkinsonism, but did not benefit from levodopa. He gradually developed severe freezing of gait (FOG) and urinary incontinence. Additionally, the patient exhibited dream-enactment behavior but denied visual hallucinations. At the time of evaluation, he was taking 200 mg of levodopa six times a day as well as chlorthalidone, cyanocobalamin and aspirin. He did not have a known family history of any similar disorders.

The patient was essentially wheelchair-bound at the time of evaluation and exhibited hypomimia and a mixed dysarthria, with hypokinetic, hyperkinetic, and ataxic components. Square wave jerks were present with visual fixation. The patient could not stand from his wheelchair without assistance. He had severe FOG, and his postural reflexes were severely impaired. The patient's legs were visibly shaking when he stood. Auscultation of the patient's quadriceps, tibialis anteriors (TAs), and medial gastrocnemii (MGs) with the bell and diaphragm of a stethoscope revealed a "thumping" sound bilaterally.

Autonomic reflex testing showed a moderate degree of generalized autonomic failure, and magnetic resonance imaging (MRI) scans of the brain showed pontocerebellar atrophy on sagittal images, findings consistent with multiple system atrophy.

SEMG revealed frequent, semi-rhythmical, 4–7 Hz bursts of motor activity lasting less than 50 ms in the bilateral TAs and MGs, greater than the quadriceps. This SEMG activity sounded like water boiling. Occasionally, these bursts fired synchronously. Subjectively, the patient felt involuntary "tightening and loosening" of his lower limb muscles (Video 1). Leaning forward onto a chair while still standing, the SEMG activity in the patient's legs became low in amplitude and tonic and lasted hundreds of milliseconds; the patient denied involuntary movements in his legs or arms at this time (Video 1). These findings are diagnostic of orthostatic myoclonus.

Based on previous studies with large patient numbers, SEMG characteristics of OM include burst durations less than 100 ms at an irregular frequency of 4–11 Hz (Fig. 1) (Glass et al., 2007; van Gerpen, 2014). OT has been demonstrated to have a rhythmical frequency of 12–18 Hz in the leg muscles of standing patients on SEMG (Fig. 2, Video 2) (McManis and Sharbrough, 1993; Piboolnurak et al., 2005). The auditory signature of OM with SEMG is similar to the sound of water boiling, unlike OT, which sounds like the rotating blades of a helicopter (Video 1 and 2) (van Gerpen, 2014).



Video 1. Demonstration of orthostatic myoclonus with accompanying SEMG. When supported, the patient's legs do not appear to shake, but the patient experiences involuntary movements in his leg muscles when standing unsupported. SEMG recordings display evidence of orthostatic myoclonus with characteristic audio signature, sounding like water boiling.

Both OM and OT cause involuntary leg movements bilaterally in standing patients (Gerschlager et al., 2004; Glass et al., 2007). OT tends to rapidly diminish when the patient begins to walk, while the lower extremity shaking observed in OM patients typically persists, impacting the patient's gait. OM is more frequently associated with concomitant with neurodegenerative disorders than OT (Gerschlager et al., 2004; Glass et al., 2007; Gasca-Salas et al., 2013; van Gerpen, 2014).

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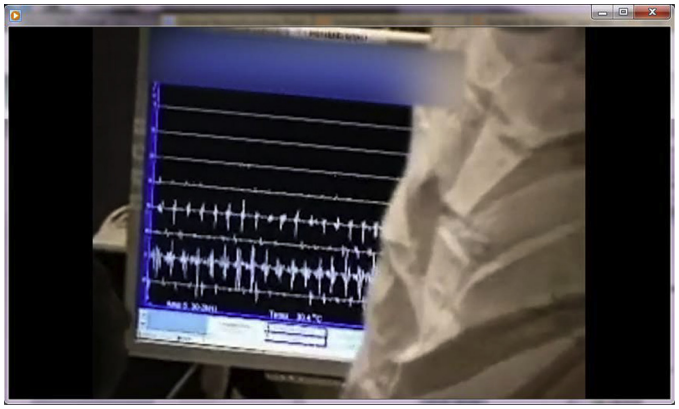

Video 2. Demonstration of orthostatic tremor with accompanying SEMG. When the patient is standing unsupported, the SEMG recording displays evidence of orthostatic tremor with characteristic audio signature of the rotating blades of a helicopter.

There have been isolated case reports that OT can be directly auscultated by means of placing the diaphragm of the stethoscope on leg muscles in standing patients (Brown, 1995; Piboolnurak et al., 2005; Yaltho and Ondo, 2014). The characteristic sound on auscultation is often described as a thumping sound (Brown, 1995; Piboolnurak et al., 2005). This has led to speculation that OT can be reliably diagnosed with auscultation and other clinical signs, obviating the need for SEMG (Brown, 1995). In fact, some authors have employed auscultation in studies as a part of their diagnostic workup of OT (Yaltho and Ondo, 2014). However, the auscultation of the leg muscles of our patient also revealed a thumping sound, yet this patient has SEMG-confirmed OM.

The findings above add further weight to the argument that SEMG is likely needed to reliably differentiate between orthostatic hyperkinesias. Auscultation of leg muscles is unlikely to be accurate in differentiating OM from OT, or even non-specific leg "shaking" while standing, the most common electrophysiological finding in a large cohort of patients with "shaky legs" (van Gerpen, 2014). A prospective study of a large cohort of patients complaining of "shaky legs", with various orthostatic hyperkinesias, comparing SEMG findings to leg auscultation may prove useful.

Disclosures

None.

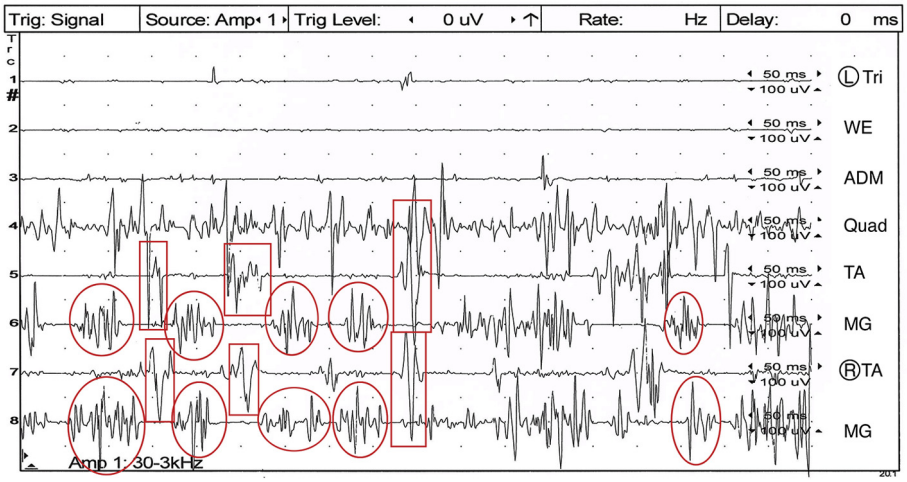

Fig. 1. SEMG tracing characteristic of OM in a standing patient. Semi-rhythmical activity between the bilateral tibialis anteriors (rectangles) and medial gastrocnemii (circles) is present.

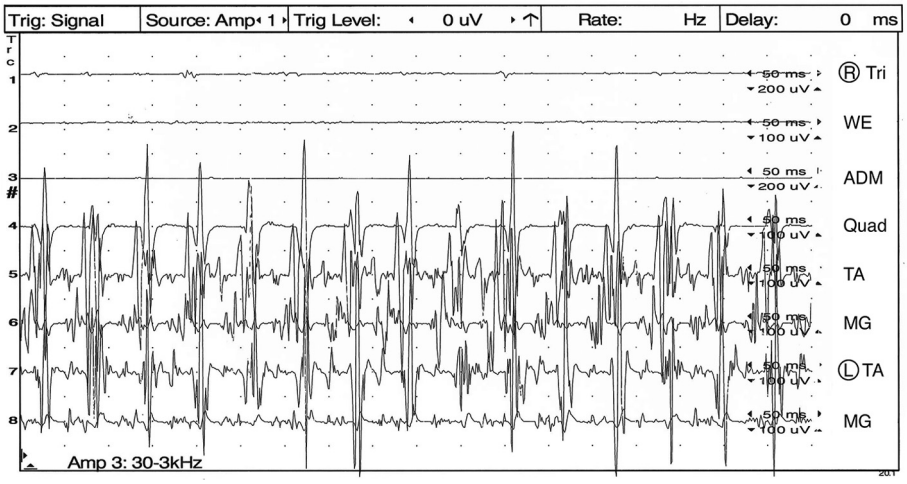

Fig. 2. SEMG tracing characteristic of OT of 16 Hz in a standing patient.

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