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Short communication

# Orthostatic myoclonus after brain tumor radiation: Insights from two lesional cases

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ABSTRACT

*Introduction:* Orthostatic myoclonus (OM) is a recognized syndrome of gait unsteadiness accompanied by lower limb myoclonus provoked by the assumption of an upright posture. OM typically affects the elderly and is often associated with neurodegenerative disease. We sought to review the clinical and electrophysiologic characteristics of OM due to brain tumor treatment, the first reported lesional cases of this rare disorder.

*Methods:* The database of the Mayo Clinic Rochester Movement Disorders Laboratory was searched for all patients diagnosed with OM from January 2007 to December 2016. All available clinical, radiographic, and surface electromyographic data were reviewed, and patients with a history of primary or metastatic brain tumor were analyzed.

*Results:* Two patients with OM and brain tumor were identified; both had undergone tumor resection and targeted brain radiation. Both patients complained of unsteadiness while walking and recurrent falls. Tumor pathology (atypical meningioma, gliosarcoma) was centered in the frontal lobe and extended to the supplementary motor area (SMA), pre-SMA, or prefrontal cortex. Medications did not improve gait.

*Conclusion:* Two cases of brain tumor-related OM suggest that degeneration of frontal motor programming circuits underlies the pathophysiology of OM.



## 1. Introduction

Orthostatic myoclonus (OM) is a recognized syndrome of gait unsteadiness accompanied by lower limb myoclonus provoked by the assumption of an upright posture [1]. The diagnosis of OM is established electrophysiologically, and 70 cases have been reported to date [1–5]. Slightly more than half of OM cases are associated with a neurodegenerative process of the central nervous system, usually Parkinson disease or multiple system atrophy. OM has never been reported in the setting of brain tumor or following brain radiation, though we recently encountered two such patients in our clinical practice.

## 2. Methods

This study was approved by the institutional review board of Mayo Clinic, Rochester, Minnesota.

### 2.1. Patient selection

We retrospectively reviewed the medical records and laboratory data of all patients diagnosed with “postural myoclonus” or “orthostatic myoclonus” in the Mayo Clinic Rochester Movement Disorders Laboratory database between January 1, 2007, and December 31, 2016. Patients were included if they had any history of primary or metastatic brain tumor.

### 2.2. Diagnosis of OM

The diagnosis of OM was based on the presence, only while standing, of irregular, high-amplitude, short-duration (less than 100 ms) muscle bursts in at least three lower limb or paraspinal muscles, at least one of which had to be a proximal muscle. The myoclonic bursts had to be present exclusively or nearly exclusively while standing and not in any other position. The presence of myoclonus at rest was exclusionary. These criteria are slightly modified (more stringent) than the original description by Glass

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et al. [1] Of 98 records reviewed, three patients had been diagnosed with OM and brain tumor, but one was excluded from further analysis because myoclonic bursts were present only in distal leg muscles.

## 3. Case descriptions

### 3.1. Patient 1

A right-handed woman was diagnosed at age 62 with a WHO grade II atypical meningioma arising from the falx and abutting the right frontal lobe. She underwent gross total resection shortly after diagnosis and then Gamma Knife radiosurgery for recurrent tumor in the superior sagittal sinus at age 63. Tumor progression prompted additional Gamma Knife radiosurgery at age 65 and then focused external beam radiation therapy (5940 cGy in 33 fractions) at age 68. Four weeks after completing brain radiation, she developed progressive imbalance and falls. The patient sensed jerking of her legs while standing that persisted while walking, resulting in a slow, cautious gait. Small-amplitude muscle twitches in the legs were visible and palpable during standing but not sitting. Her neurologic exam otherwise revealed left hyperreflexia, marked distal pyramidal weakness, and Babinski sign, residua of her original tumor surgery, without parkinsonism or other evidence of a progressive neurodegenerative syndrome.

Three months after symptom onset, multichannel surface EMG showed high-amplitude short-duration muscle bursts (50–120 msec) predominantly in tibialis anterior and less frequently quadriceps and hamstrings while standing. These muscle bursts appeared in the lumbar paraspinal muscles while leaning forward with weight supported on her arms but were completely absent while sitting, diagnostic of orthostatic myoclonus (Fig. 1). MRI brain scan one month and again twelve months after onset of imbalance showed right frontal encephalomalacia with stable residual tumor invading the superior sagittal sinus and falx (Fig. 3A). Potentially confounding medications included citalopram and verapamil at the start of gait unsteadiness and OM diagnosis, but these were not thought to be causative of her syndrome because she had been on both medications for at least 3 years prior to any gait difficulties and because discontinuation of citalopram did not improve her gait. Treatment with clonazepam, levetiracetam, and carbidopa/levodopa were all ineffective during 30 months of follow up.

### 3.2. Patient 2

A right-handed woman was diagnosed at age 63 with a WHO grade IV gliosarcoma in the right frontal lobe. She underwent gross total resection, focused external beam radiation therapy (dose

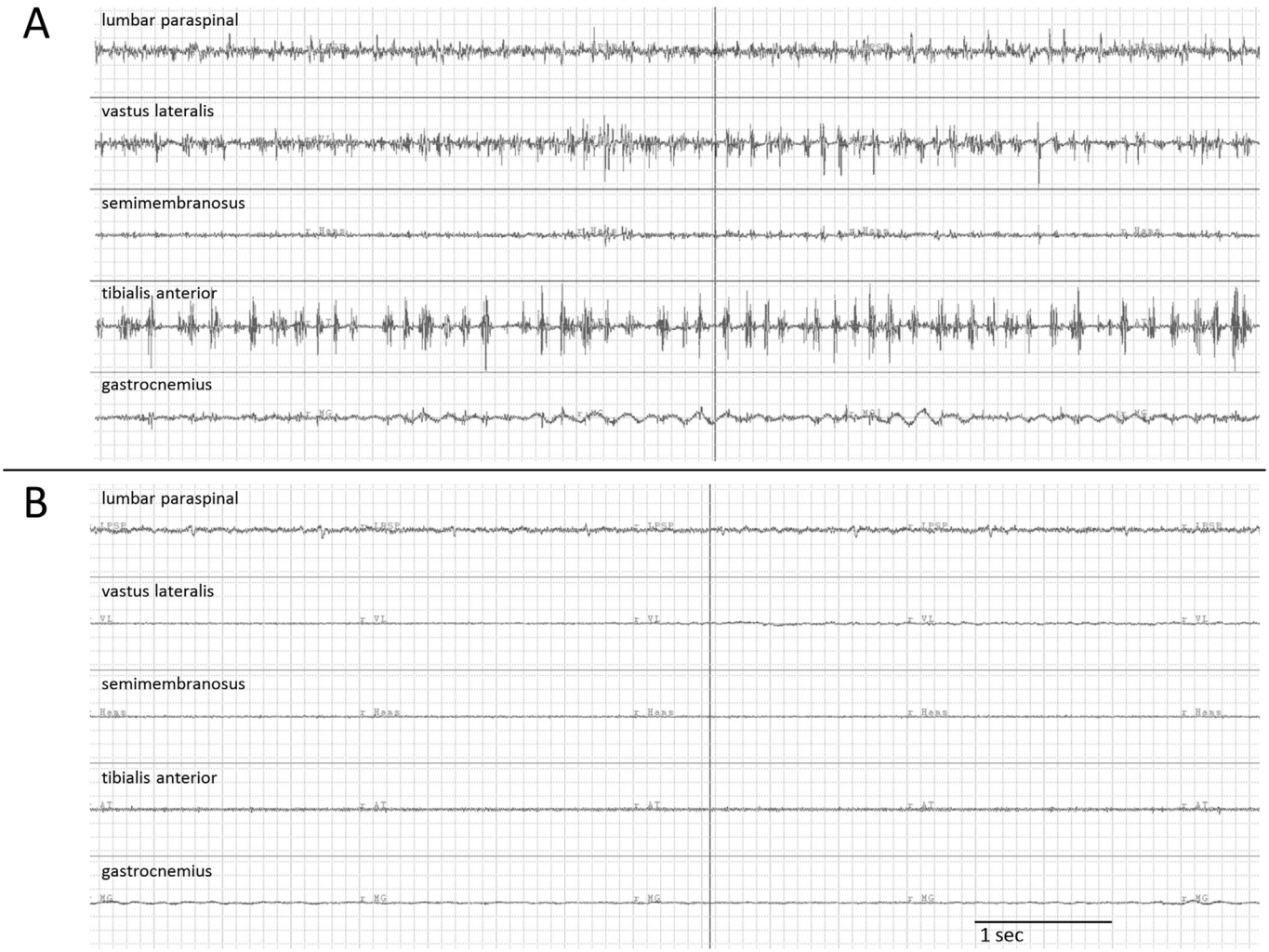


**Fig. 1.** Multichannel surface EMG in patient 1 shows myoclonic bursts in hamstrings, quadriceps, and tibialis anterior muscles while standing (A) but not while sitting (B).

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