Limb myorhythmia treated with chemodenervation: a case report

Ther Adv Neurol Disord

2023, Vol. 16: 1–5

DOI: 10.1177/17562864221150317

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***Abstract*:** We describe a case of limb myorhythmia successfully palliated with botulinum toxin injections. The patient is a 30-year-old male evaluated for abnormal movements of the left lower foot that began after an ankle injury for which the patient underwent Achilles tendon scar tissue debridement without improvement. On examination, he had near-constant involuntary, slow, rhythmic flexion/extension tremor of toes 2–4 that was diminished during active movement. Needle electromyography (EMG) revealed a rhythmic, 2–3 Hz tremor isolated to the flexor digitorum brevis. After failure of medical management with muscle relaxants, gabapentin, and levodopa trials, the patient underwent two EMG-guided chemodenervation procedures with incobotulinum toxin A injections of the left flexor digitorum brevis. At 3-month follow-up, he had achieved a sustained 50% reduction in the intensity of the movements and improved quality of life. Myorhythmia is a rare condition characterized by a repetitive, rhythmic, slow frequency (1–4 Hz) movement affecting the cranial and limb muscles. The most common causes include stroke, demyelinating disorders, drug or toxin intake, trauma, and infections. The management of this condition is very limited with pharmacologic agents such as anticholinergics, antispasmodics, anticonvulsants, or dopaminergic agents showing limited efficacy. The use of botulinum toxin chemodenervation aided by EMG muscle targeting can be a useful therapeutic intervention in cases of medication-refractory regionally distributed myorhythmia involving accessible muscles.

Keywords: botulinum toxin, case report, chemodenervation, myorhythmia

Received: 5 August 2022; revised manuscript accepted: 21 December 2022.

Case report

We describe a case of limb myorhythmia treated with botulinum toxin injection. The patient is a 30-year-old male first seen in neurology clinic on 1 April 2019 for abnormal movements of his left leg which started in 2016. He described them as visible movement of the toes of his left foot with associated pulsation and twitching of the lower medial and posterior leg at the level of the Achilles tendon. The patient endorsed that the movements were constant, present at rest and diminished with ambulation, with no urging and no other alleviating or exaggerating factors. The abnormal movements involving his toes were mildly painful and occasionally caused him difficulty falling asleep. He denied sleep awakenings because of these symptoms. Remarkably, the patient was an ultra-marathon runner and had a prior history of a left ankle sprain and Achilles tendinopathy diagnosed in 2016, for which he underwent Achilles scar tissue debridement surgery and a gastrocnemius resection procedure with no significant improvement in his tendinitis symptoms. Following this injury and subsequent surgical procedure, the patient developed the described left leg tremor.

On evaluation, the patient neurological examination was significant for a very slow repetitive rhythmic flexion/extension oscillatory movement of toes 2–4 in the left foot with occasional abduction that was distractible and suppressible for a short period of time. The movements decreased in amplitude with active motion. The patient also exhibited a near-constant involuntary intermittent twitching of the medial inframalleolar area.

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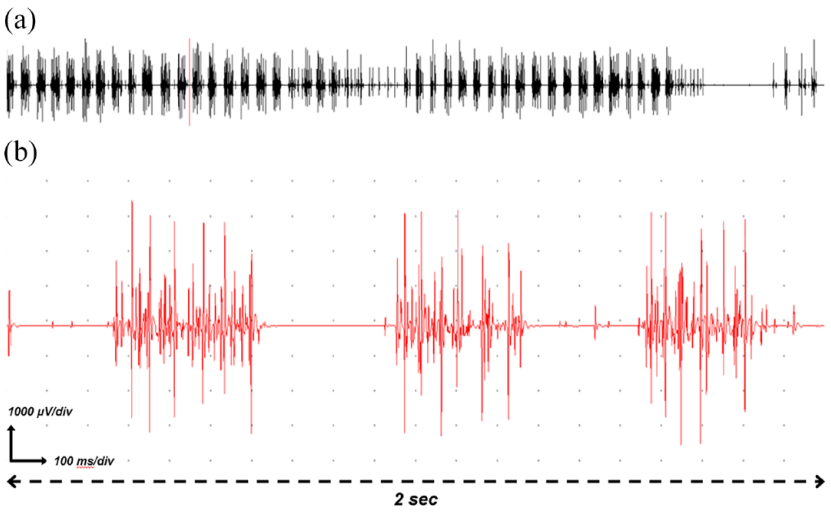

Figure 1. EMG of the patient left flexor digitorum brevis muscle. Rhythmic contractions at a frequency of 2–3 Hz can be appreciated in the tracing. (a) General view of the patient's EMG tracing. (b) Close-up view of the EMG tracing.

There were no associated sensory or strength deficits, appendicular or central ataxia, upper or lower motor neuron signs. Supplemental Figure 1 (Video) presents the features of the patient left toe movements. Needle electromyography (EMG) examination of selected muscles in the left lower extremity was also performed, which revealed a rhythmic slow 2–3 Hz tremor isolated to the flexor digitorum brevis muscle (Figure 1). Magnetic resonance imaging (MRI) of the left ankle only revealed a noninsertional Achilles tendinosis and a posterior tibialis tendon sheath effusion. A brain MRI found no evidence of intracranial abnormalities, infarction, hemorrhage, or extra-axial fluid collections. MRI of the cervical and thoracic spine only showed a mild foraminal stenosis at the C3–C6 levels, minimal central canal stenosis, and small disk protrusions between T6–T7 and T7–T8.

With this evidence, a diagnosis of limb myorhythmia was made. For treatment, the patient was initially trialed on symptomatic management with muscle relaxants and gabapentin 200 mg three times daily, achieving no relief in his presenting symptoms. This was followed by a 2-month trial of carbidopa/levodopa therapy up to a maximum dose of carbidopa 50 mg/levodopa 200 mg that was also unsuccessful. Given his presentation was refractory to medication management, on 20 July 2021 the patient underwent an EMG-guided chemodenervation procedure with incobotulinum toxin A 40U injection on the left flexor digitorum brevis. Figure 2 presents a diagram of the injection site. He tolerated the procedure well without immediate complications. At 3-month follow-up, the patient had achieved a long-standing 40–50% decrease in the intensity of the myorhythmia. On repeat evaluation, he only described a residual twitching posterior to the navicular bone and a dull pain of 1/10 intensity in the tibialis posterior tendon upon plantar flexion of the first digit. Given this improvement, on 30 November 2021 the patient underwent a second flexor digitorum brevis muscle EMG-guided incobotulinum toxin A 50U injection, again tolerating the procedure well with no complications. On his subsequent 3-month follow-up visit, the patient noted a sustained, greater than 50%, improvement in the amplitude of the movement. The patient also endorsed having an improved quality of life and a resolution of his initial insomnia due to discomfort from the movements.

Discussion

Myorhythmia is defined as a repetitive, rhythmic, slow frequency (1–4 Hz) movement[1] affecting mainly the cranial and limb muscles. The tremor typically occurs at rest but can occasionally be present with movement and voluntary activity, usually disappearing with sleep. It is caused by an

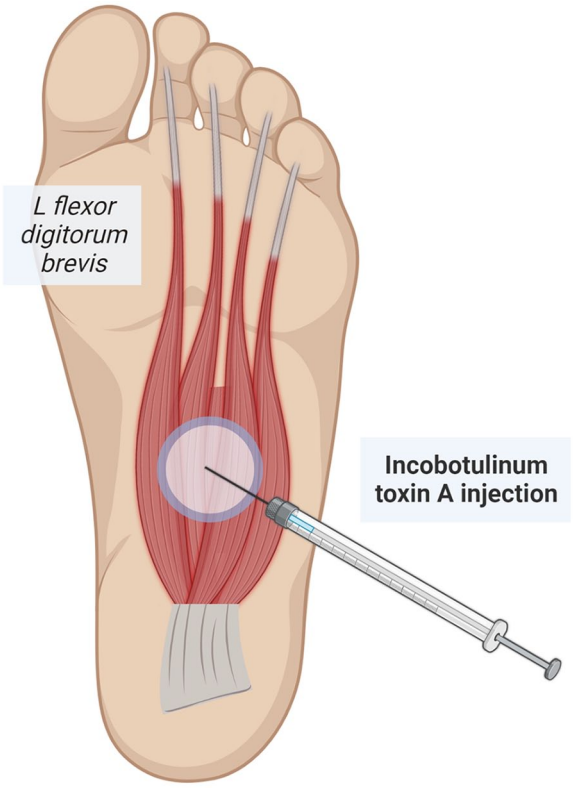

Figure 2. Incobotulinum toxin A injection site at the left flexor digitorum brevis muscle.

alternating or synchronous contraction of antagonist muscles. Needle EMG typically shows discharges of motor units with normal morphology and a slow inter-burst rate of 1–4 Hz. This condition needs to be distinguished from tremor dominant parkinsonism. The main difference between these is the slower rate (2–3 Hz) of movement on EMG in myorhythmia compared with the 4–6 Hz of parkinsonian tremor.[2] Other differential diagnoses include dystonic tremor, Holmes tremor, cortical tremor, and myokymia.[1] In our patient case, the slowness of the rate and rhythmicity of the tremor on physical examination along with the EMG findings gave the diagnosis of myorhythmia.

The most common cause of myorhythmia is brainstem[3] and thalamic strokes, but it can also be found in patients with autoimmune[4,5] and demyelinating disorders, drug or toxin intake, trauma, *Listeria* rhombencephalitis[6] and Whipple disease where it commonly manifests with oculomasticatory rather than limb myorhythmia.[7] Case reports have also described myorhythmia associated with COVID-19 Acute Hemorrhagic Necrotizing Encephalopathy,[8] brain tumors (i.e. spinal cord glioma),[9] Wilson disease[10] Hodgkin's lymphoma[11] and anti-IgLON5 disease.[12] Interestingly, our patient only had a history of Achilles tendonitis for which he had undergone surgical interventions, but the rest of his medical and family history was noncontributory.

The underlying substrate in limb myorhythmia has been studied in electrophysiological studies,[13] which have revealed cerebellum and brainstem dysfunction at the lateral olivopontomedullary level. Imaging and autopsy evaluations have also shown unilateral[14,15] hypertrophic inferior olivary degeneration in these patients. Lesions in other components of the Guillain–Mollaret triangle (i.e. contralateral dentate nucleus, red nucleus and superior cerebellar peduncle) and the bilateral substantia nigra have also been described.[2] In our patient case, brain and spine MRI imaging studies did not reveal any remarkable findings.

Overall, the close temporal and anatomical relationship between the lower extremity injury and the development of myorhythmia is suggestive of

a possible association between these two events. The absence of other risk factors, systemic symptoms or structural abnormalities, and the stable disease course also make other etiologies of myorhythmia less likely. Trauma to the peripheral nervous system is a well-recognized cause of dystonia,[16,17] tremor, myoclonus, spasms, and even parkinsonism.[18,19] It is presumed that cortical and subcortical reorganization as a response to altered sensory input secondary to a peripheral lesion could be the trigger for these abnormal movements.[19] In line with this prior evidence, here we presented the first case report of a patient with myorhythmia following a traumatic peripheral injury.

The management of myorhythmia is very limited. Pharmacologic approaches with anticholinergics, antispasmodics, anticonvulsants, levodopa, and dopamine antagonists have shown limited to no efficacy. Deep brain stimulation of the thalamus was also trialed in a patient with refractory myorhythmia secondary to a pontomesencephalic hypertensive hematoma, without improvement in his symptoms.[20] Botulinum toxin acts by binding to cholinergic nerve terminals presynaptically and decreasing the release of acetylcholine as a result. This leads to a persistent muscle relaxation effect that could potentially help prevent the intermittent contraction of the affected muscles in patients with regionally distributed myorhythmia. Palatal tremor (formerly palatal myoclonus) is a condition within the spectrum of myorhythmia that typically presents with tinnitus secondary to rhythmic involuntary movements of the soft palate and is often refractory to medical management. Five patients with palatal myoclonus were treated with botulinum toxin injections of the levator and tensor veli palatini muscles. Of the five, four achieved complete resolution of symptoms after a course of treatment and only one reported transient side effects.[21] Another case report described the benefit of botulinum toxin injections of the left orbicularis oris and depressor anguli oris muscles in a patient with peribuccal and pharyngeal myorhythmia.[15] Despite this evidence, there have not been any previous reports on the use and efficacy of botulinum toxin injections in patients with limb myorhythmia. Here we described the case of a 30-year-old patient who achieved a partial resolution of his presenting left lower extremity motor symptoms with the use of this chemodenervation procedure.

Declarations

Ethics approval and consent to participate

Our study did not require an ethical board approval because it constitutes a case report. A written consent form was obtained from the patient in which patient voluntarily consented without compensation to participate in this case report.

Consent for publication

A written consent form was obtained from the patient in which he voluntarily consented without compensation to the use of his case and video/audio recording of his abnormal movements for education purposes including publication in a journal.

Author contributions

Nil Saez-Calveras: Visualization; Writing – original draft.

Meredith Bryarly: Investigation; Resources.

Meagen Salinas: Conceptualization; Supervision; Writing – review & editing.

Acknowledgements

We deeply thank our patient for his willingness to participate in this case report.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Competing interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Availability of data and materials

Data sharing not applicable to this article as no data sets were generated or analyzed during the current study. However, the patient's deidentified EMG recording is available from the authors upon reasonable request and with permission of the patient.

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Supplemental material

Supplemental material for this article is available online.

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