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Case report

Adult-onset primary focal foot dystonia

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Abstract

Although primary focal hand dystonia has been well characterized, primary focal foot dystonia in adults has rarely been reported. Our objective was to describe the clinical phenotype and treatment outcomes in patients with primary, adult-onset focal foot dystonia. To this end we conducted a retrospective study of four consecutive patients (59.5 ± 13.5 years, range 44–67) diagnosed over a period of 6 years and followed-up for at least 5 years. Focal foot dystonia resulted in variable physical impairment. Anti-dystonic agents were mildly effective whereas botulinum toxin injections provided substantial benefit when used. Focal primary focal foot dystonia seems to be a rare form of focal dystonia with distinct clinical features that may benefit from treatment with botulinum toxin.

Keywords: Dystonia; Focal; Foot; Adult-onset

1. Introduction

Of the focal limb dystonias, primary focal hand dystonia has been well studied since its first description by Sheehy and Marsden [1]. On the other hand, primary focal foot dystonia in adults has rarely been reported. To better clarify the characteristics of this rare form of focal dystonia we conducted a retrospective study of four consecutive patients seen in our department.

2. Methods

All patients were diagnosed (over a period of 6 years) and followed-up at the Movement Disorders Clinic of the Department of Neurology of University of Miami, School of Medicine. None of our patients had a family history of dystonia or other extrapyramidal disease. Only one patient had history of other neurological disease (case 3, migraine). None had radiological (CT or MRI) evidence of basal ganglia lesions. There was no history of weakness, injury or surgery involving the foot or leg predating the movement disorder. Dystonia was action induced and remained focal during follow-up (at least 5 years). None of our patients developed other extrapyramidal symptomatology. A global impression scale was used in the evaluation of the therapeutic outcomes (0 = no effect, 1 = mild, 2 = moderate, 3 = marked). The characteristics of our patients are presented in Table 1. The study was approved by the local Institutional Review Board (IRB).

3. Results

3.1. Case 1

This 68-year-old right-handed female was initially evaluated for involuntary movements of the left foot. The patient had been symptomatic for the preceding year with progressive inversion and plantar flexion of the left foot. Initially, symptoms were elicited by walking but gradually evolved to being produced by standing. She had also started experiencing occasional spontaneous foot inversions while seated. There were no other associated symptoms. Brain MRI was unremarkable. Spine MRI only showed mild degenerative changes.

Neurological examination was normal except for moderate inversion of the left foot with hyperflexion of the toes (including the big toe) during walking with mild

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Table 1
Demographic and clinical characteristics of patients with primary focal foot dystonia

Case	Sex	Onset age	Limb affected	Follow-up (years)	Characteristics	Impairment	Pain	Medication trials	BTX	Response to treatment
#1	F	67	Left	5	Foot inversion, toe flexion	Mild	No	L-dopa	Yes	Marked (BTX)
#2	F	74	Left	8	Foot inversion and plantar flexion, toe flexion	Moderate	No	L-dopa, Cy, Bx	Yes	Marked (BTX)
#3	F	44	Left	6	Foot inversion, toe splaying	Mild	Yes	L-dopa, Ba, Tr, Cl, Ga	No	Moderate (oral medications)
#4	M	53	Left	5	Ankle and toe dorsiflexion	Very mild	Yes	L-dopa, Ba, Am,	No	Mild (oral medications)
Koller [4]	M	50	Right	N/A	Flexion ankle and toe, foot inversion	Moderate	No	Tr	No	Moderate
Duarte [6]	M	64	Right	1.5	Foot inversion and dorsiflexion	Moderate	Yes	L-dopa	Yes	Marked
Kim [5]	F	46	Left	1	Foot eversion, toe flexion	Mild	No	L-dopa, Ba, Tr, phenol injections	Yes	Marked[a]

Ba, baclofen; Am, amantadine; Cy, cyclobenzaprine; Bx, benzexhole; Tr, trihexyphenidyl; Cl, clonazepam; Ga, gabapentin; BTX, botulinum toxin type A.
[a] Improvement was due to intramuscular phenol injections and not botulinum toxin injections.

functional impairment. Symptoms were milder in backward walking and subsided when the patient stopped or rested.

After an initial unsuccessful trial of carbidopa/levodopa and diazepam, the patient received botulinum toxin (BTX) injections under EMG guidance in the left flexor digitorum longus (up to 50 U of BOTOX®) and the left flexor hallucis longus (up to 50 U). Initially, she experienced improvement in ambulation with a decrease in toe hyperflexion. The patient subsequently received injections in the left gastrocnemius (up to 50 U) and the left tibialis posterior (up to 50 U) with moderate benefit.

3.2. Case 2

This 75-year-old right-handed female was initially evaluated for involuntary posturing of her left foot. One year prior to presentation she started experiencing mild flexion of her left toes which progressed over time and eventually resulted in a combination of toe flexion, foot inversion and plantar flexion. Symptoms were triggered by ambulation and subsided during rest and resulted in significant disability, as she became unable to walk long distances. She denied any other neurologic deficits.

On examination she exhibited flexion of the toes associated with plantar flexion and inversion of the left foot after the first few steps of ambulation. Symptoms became more prominent with every successive step, limiting ambulation. During backward motion, there was significant diminution of the abnormal posturing. Neurological examination was otherwise unremarkable.

After unsuccessful trials of L-dopa, cyclobenzaprine and benzhexole, the patient started receiving BTX injections under EMG guidance. She received injections (32 sessions) in the left flexor digitorum longus (up to 100 U), the left flexor hallucis longus (up to 50 U), the left flexor digitorum longus (up to 100 U), the left flexor digitorum brevis (up to 25 U), and the left tibialis posterior (up to 100 U). The quality of response to the injections has varied from marked (90% improvement over a 2.5-month period) to mild (20% improvement over a period of 1.5 months). She continues being followed with quarterly sessions of botulinum toxin injections for the last 10 years.

3.3. Case 3

This 45-year-old right-handed female was initially evaluated for involuntary movements that consisted of inversion of the left foot and toe splaying associated with marked left calf pain. Symptoms progressed for a year until they became very frequent (every 5–30 min), usually triggered by emotion, exhaustion, walking, or some other exertion. Spinal MRI revealed mild degenerative changes. Brain MRI, EMG, nerve conduction and copper studies were normal. On examination foot tapping and walking were initially normal but within seconds triggered inversion of the left foot which persisted for as long as she would continue ambulating. Even while seated, there was a tendency for the left foot to spontaneously invert.

She was initially placed on carbidopa/levodopa and baclofen with limited success. After tapering carbidopa/levodopa, baclofen remained the mainstay of the antidystonic regimen with subsequent trials of adding trihexyphenidyl, clonazepam and gabapentin. The patient (7 years after the diagnosis) remains on a combination of baclofen and clonazepam which seems to provide her with a measure of relief. There has not been much in terms of change or of her focal dystonia. She has declined repeated offers of botulinum toxin injections.