CASE REPORT

Adult onset primary focal dystonia of the foot: an orthopaedic intervention

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SUMMARY

A 54-year-old woman presented to a foot centre with a chief symptom of cramping in her toes, which, she believed, was of a secondary cause originating from a bunion. She was treated conservatively; however, she returned a month later as the symptoms had progressed to painful cramping of toes, toe-curling and instability while walking, due to involuntary movement of her toes. It was believed that the patient presented with a rare case of primary adult onset focal foot dystonia. This case report explains dystonia further in detail and delves into the different treatment and management options available today, including the unique orthopaedic intervention provided for this patient.

BACKGROUND

Dystonia is a movement disorder characterised by involuntary muscle contractions that cause slow repetitive movements or abnormal postures. Although childhood-onset dystonia is usually primary in nature and frequently begins in the foot and spreads to other parts of the body, adult-onset dystonia that initially presents in the foot is rare, generally does not spread and is usually secondary to another condition. Aetiologies of adult onset foot dystonia include Parkinsonism, stroke, trauma, a structural lesion and psychogenic dystonia.¹⁻³ Other less common causes are Wilson's disease, exposure to certain medications and spinal stenosis.

There is no gold standard treatment for dystonia but the condition can be managed in a variety of ways. The most popular treatment for adult onset focal dystonia is botulinum toxin.^{1 2 4 5} The toxin can be targeted to specific muscles that are contracted and cause involuntary movement. It does not work in every patient and other medications are sometimes utilised such as anticholinergics, GABAergic and dopaminergic agents.⁶ Physical therapy can also be helpful in management of the contracting muscles via stretching exercises, splinting, ankle-foot-orthosis, functional electrical stimulation or deep brain stimulation.⁷⁻⁹

The participant in our case report demonstrates an adult onset primary focal dystonia of the left foot. To date, no other secondary condition has been diagnosed to explain her dystonia. However, the development of a neurodegenerative condition at a later date cannot be ruled out.

CASE PRESENTATION

A 54-year-old woman presented to a foot centre with a chief symptom of a 'cramping sensation of the left big toe that travels to the smaller toes'. The

Figure 1 Involuntary movements of the patient's toes in L foot. She described her toes as 'curling and cramping' involuntarily, which progressively worsened.

patient stated that the pain and cramping began 4 months earlier and she was not sure if it was related to her left 'bunion'. The patient stated that when she walked she felt a 'grabbing pain' that seemed to be related to her 'toes curling'. The patient denied any numbness or burning pain of the feet or legs. She also denied any involuntary movement of other parts of her body. Her medical history was unremarkable and negative for the use of neuroleptic or dopaminergic medications. There was no history of trauma to the head, back or legs. There was no family history of neurological disorders.

One month after initial presentation, the patient stated that her left big toe had 'started to go down' when she walked and the pain level of her left big toe had increased from a level of 4 to a level of 7 of 10. The patient stated that her toes were 'curling and cramping' so much that she could not fit into a closed-toed shoe and she had had to start using a cane when walking. Three months after her initial presentation, the patient stated that she was now

Video 1 Dystonia of L foot while the patient is sitting.

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Video 2 Patient's gait where the intermittent supination of the L foot with persistent movements of the digits can be noticed.

walking on the 'outside of her left foot' and that the toes of her left foot kept "moving by themselves and cramping" (figure 1).

INVESTIGATIONS

Physical examination revealed intermittent flexion, extension and fanning of the digits of the left foot (video 1). In gait, intermittent supination of the left foot was noted with persistent

movement of the digits of the left foot (video 2). No abnormalities of deep tendon reflexes, muscle power or sensation were noted. No pain was elicited with a straight leg raise. No wasting or fasciculation of muscles was evident. Cogwheel rigidity of the arm, tremor and facial hypomimia were absent.

Utilising an F-scan by Tekscan, a computer assisted in-shoe kinetic gait analysis system, we evaluated the patient's gait changes when employing a gauntlet style lace-up ankle brace, a digital crest applied to the shoe's innersoles, and a shoe with a mild rocking platform. Our goal was to see what item or combination of items provided improved stability and overall postural symmetry. A baseline evaluation with the patient's own running shoes was compared to the ankle brace, digital crest, rocker shoe and various combinations of these items. Both, the active propulsive phase as well as the single limb support phases were examined and compared.

When the base study was sequentially examined as the additional items were included, it was noted that while selective improvements in either the active propulsive phase, or single limb support were individually improved, the most notable improvement was recorded when the digital crest and ankle wrap were used with the shoe that had a mild rocker platform (figures 2–6).

DIFFERENTIAL DIAGNOSIS

Further analysis by a neurologist included MRI of the brain and spine, electromyography analysis and complete haematological

Figure 2 Evaluation of phases of gait using F-scan. Baseline results of the patient in running shoes.

Parameter	Normal Range	Left Foot			Right Foot		
		Avg	Variation	StdDev	Avg	Variation	StdDev
Contact	14 - 20	17	16 - 19	1	19	17 - 21	1
Foot Flat	16 - 22	54	31 - 65	12	59	47 - 67	6
Midstance	29 - 37	58	54 - 62	3	48	42 - 55	4
Propulsive	45 - 55	25	21 - 30	3	33	24 - 39	5
Active Propulsive	31 - 35	5	0 - 11	4	15	6 - 22	5
Passive Propulsive	14 - 20	20	18 - 22	1	18	17 - 19	1
Single Support	60 - 72	63	59 - 65	2	63	61 - 66	2
Total Double Support	28 - 40	37	35 - 41	2	37	34 - 39	2
Initial Double Support	14 - 20	17	16 - 19	1	19	17 - 21	1
Terminal Double Support	14 - 20	20	18 - 22	1	18	17 - 19	1

Note: Red entries indicate an average value that is outside the Normal Range

Figure 3 Results of the patient in a digital crest pad.

Parameter	Normal Range	Left Foot			Right Foot		
		Avg	Variation	StdDev	Avg	Variation	StdDev
Contact	14 - 20	18	17 - 20	1	20	18 - 21	1
Foot Flat	16 - 22	57	53 - 61	4	60	49 - 68	8
Midstance	29 - 37	57	54 - 58	2	49	43 - 59	5
Propulsive	45 - 55	26	24 - 28	1	31	22 - 37	6
Active Propulsive	31 - 35	6	3 - 8	2	14	2 - 20	6
Passive Propulsive	14 - 20	20	19 - 21	1	18	16 - 20	1
Single Support	60 - 72	62	60 - 64	2	62	61 - 63	1
Total Double Support	28 - 40	38	36 - 40	2	38	37 - 39	1
Initial Double Support	14 - 20	18	17 - 20	1	20	18 - 21	1
Terminal Double Support	14 - 20	20	19 - 21	1	18	16 - 20	1

Note: Red entries indicate an average value that is outside the Normal Range

Figure 4 Patient in digital crest pad and gauntlet style ankle brace.

Parameter	Normal Range	Left Foot			Right Foot		
		Avg	Variation	StdDev	Avg	Variation	StdDev
Contact	14 - 20	18	17 - 20	1	18	16 - 19	1
Foot Flat	16 - 22	54	43 - 65	8	59	53 - 68	6
Midstance	29 - 37	58	57 - 59	0	52	42 - 67	8
Propulsive	45 - 55	23	22 - 25	1	30	16 - 39	7
Active Propulsive	31 - 35	5	3 - 6	1	9	15 - 21	12
Passive Propulsive	14 - 20	18	18 - 19	1	18	16 - 21	2
Single Support	60 - 72	63	61 - 65	1	61	51 - 65	5
Total Double Support	28 - 40	37	35 - 39	1	39	35 - 49	5
Initial Double Support	14 - 20	18	17 - 20	1	18	16 - 19	1
Terminal Double Support	14 - 20	18	18 - 19	1	21	18 - 33	5

Note: Red entries indicate an average value that is outside the Normal Range

and biochemical analysis including copper levels. To date, no secondary cause of the foot dystonia has been established.

TREATMENT

Trihexyphenidyl was prescribed for a short period of time with no improvement of symptoms. Baclofen is currently being taken by the patient but she reports of marked fatigue with use of the medication. The following video demonstrates the result of the patient's dystonia after taking Baclofen for 1 month. There is a

visual improvement in the dystonic movement of the foot compared to pre-Baclofen therapy (video 3). Botulinum toxin has also been recommended but approval by her insurance is pending.

The patient was also prescribed a gauntlet style ankle-stabilising orthosis with strapping that limits plantar flexion and inversion of the foot (figure 7). The patient reported improved stability in gait and less difficulty in wearing a closed shoe when using the ankle brace.

Figure 5 Patient in ankle brace and rocker-bottom shoe.

Parameter	Normal Range	Left Foot			Right Foot		
		Avg	Variation	StdDev	Avg	Variation	StdDev
Contact	14 - 20	12	11 - 14	1	16	14 - 19	2
Foot Flat	16 - 22	?	? - ?	0	58	48 - 67	7
Midstance	29 - 37	44	32 - 65	12	56	55 - 58	2
Propulsive	45 - 55	44	21 - 57	12	28	22 - 31	4
Active Propulsive	31 - 35	25	-6 - 38	17	15	6 - 19	6
Passive Propulsive	14 - 20	17	15 - 21	3	13	11 - 17	2
Single Support	60 - 72	69	58 - 74	6	71	64 - 75	4
Total Double Support	28 - 40	31	26 - 42	6	29	25 - 36	4
Initial Double Support	14 - 20	12	11 - 14	1	16	14 - 19	2
Terminal Double Support	14 - 20	18	15 - 28	5	13	11 - 17	2

Note: Red entries indicate an average value that is outside the Normal Range

Figure 6 Patient in digital crest pad, ankle brace and rocker bottom shoe.

Parameter	Normal Range	Left Foot			Right Foot		
		Avg	Variation	StdDev	Avg	Variation	StdDev
Contact	14 - 20	12	11 - 14	1	18	18 - 19	0
Foot Flat	16 - 22	84	79 - 92	5	47	37 - 62	10
Midstance	29 - 37	49	44 - 56	4	49	42 - 53	4
Propulsive	45 - 55	39	33 - 44	4	32	29 - 39	4
Active Propulsive	31 - 35	20	11 - 26	6	19	15 - 27	5
Passive Propulsive	14 - 20	19	18 - 22	2	13	11 - 15	2
Single Support	60 - 72	69	67 - 71	2	69	67 - 70	1
Total Double Support	28 - 40	31	29 - 33	2	31	30 - 33	1
Initial Double Support	14 - 20	12	11 - 14	1	18	18 - 19	0
Terminal Double Support	14 - 20	19	18 - 22	2	13	11 - 15	2

Note: Red entries indicate an average value that is outside the Normal Range

Video 3 Patient on Baclofen therapy for 1 month. There is a visual reduction in dystonic movements compared to the initial visit.

OUTCOME AND FOLLOW-UP

In this particular study, the F-scan results demonstrated that not only did the patient achieve a bilateral double-digit active propulsive phase—symmetry of single limb support was also achieved. Although the combination of digital crest, gauntlet-style ankle brace and rocker bottom shoe showed the most improvement objectively, the patient was not comfortable with the use of the digital crest or rocker-bottom shoe.

Subjectively, the patient was most comfortable (VAS pain scale reduced to a pain level of 2) with the use of the gauntlet style ankle stabilising orthosis alone. Objectively, the patient's dystonia was visibly reduced after taking Baclofen for 1 month. The change in gait parameters with different modifications (digital crest, ankle brace and rocker bottom shoe) also demonstrated improvement in the patient's gait and reduction in dystonia. Currently, she is only wearing the ankle orthosis. She is still pending approval from her insurance provider for botulinum toxin therapy as it has shown good results in other studies of patients with dystonia.^{1 3–5}

DISCUSSION

Dystonic posturing of the foot in childhood is a typical presentation of a primary dystonia and often progresses to a generalised dystonia. Adult-onset foot dystonia is generally secondary in nature and is associated with Parkinson's disease,¹ trauma² and other conditions. It is important to recognise adult onset foot dystonia and to rule out secondary causes. MR of the central neuroaxis is critical in ruling out structural lesions. Some patients should have a trial of levodopa as this can act as a diagnostic aid.² Failure to diagnose foot dystonia may lead patients to undergo batteries of tests and unnecessary therapies, including surgery.¹⁰

Our case is one of those rare cases of primary adult onset foot dystonia. Studies of patients with this rare condition suggest that it generally does not spread to other areas of the body and that botulinum toxin may be of substantial benefit for some patients.^{1 3–5} The simple orthopaedic splint recommended for this patient provided reduction of pain, improved ambulation and the ability to wear closed-toed shoe gear rather than unstable sandals. This simple orthopaedic intervention may

Figure 7 Gauntlet style ankle stabilising orthosis with strapping that limits plantar flexion and inversion of the foot. The patient stated that it had improved her stability.

serve as a treatment option for other patients with this condition.

Learning points

- ▶ Dystonia is a movement disorder characterised by involuntary muscle contractures that cause slow repetitive movements or abnormal postures.
- ▶ Adult onset foot dystonia is generally secondary in nature and is associated with Parkinson's disease, trauma, stroke and other conditions.
- ▶ There is no gold standard treatment for dystonia, but it can be managed in a variety of ways. A popular treatment option for focal foot dystonia is injections of botulinum toxin.
- ▶ Simple orthopaedic intervention with braces can help manage the condition and lead to a more comfortable life.

Contributors LL is the podiatric doctor who initially saw this patient and has been following up with her. BR is the podiatric doctor who the patient was referred to by LL and who has helped with the treatment of this patient. MSD is a rising fourth-year podiatry student. She was the student doctor who initially saw and followed this patient through her course of treatment.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Schneider SA, Edwards MJ, Grill SE, *et al.* Adult-onset primary lower limb dystonia. *Mov Disord* 2006;21:767–71.
- 2 McKeon A, Matsumoto J, Bower J, *et al.* The spectrum of disorders presenting as adult-onset focal lower extremity dystonia. *Parkinsonism Relat Disord* 2008;14:613–19.
- 3 Schaller G, Tai S, Valavanis A, *et al.* Traumatic dystonia: an important orthopaedic differential diagnosis. *Foot Ankle Int* 2014;35:623–5.
- 4 Singer C, Papapetropoulos S. Adult-onset primary focal foot dystonia. *Parkinsonism Relat Disord* 2006;12:57–60.
- 5 Ramdhani RA, Frucht SJ. Adult-onset idiopathic focal lower extremity dystonia: a rare task-specific dystonia. *Tremor Other Hyperkinet Mov (N Y)* 2013;3:pii: tre-03-142-2990-1
- 6 Chang FC, Josephs KA. Levodopa responsiveness in adult-onset lower limb dystonia is associated with the development of Parkinson's disease. *Tremor Other Hyperkinet Mov (N Y)* 2013;3:pii:tre-03-150-3598-2.

- 7 Barrett MJ, Bressman SB, Levy OA, *et al*. Functional electrical stimulation for the treatment of lower extremity dystonia. *Parkinsonism Relat Disord* 2012;18:660–1.
- 8 Ostrem JL, Markun LC. Effect of frequency on subthalamic nucleus deep brain stimulation in primary dystonia. *Parkinsonism Relat Disord* 2014;20:432–8.
- 9 Schrader C, Capelle HH, Kinfe TM. GPI-DBS may induce a hypokinetic gait disorder with freezing of gait in patients with dystonia. *Neurology* 2011;77:483–8.
- 10 Schott GD. The idiopathic dystonias. *J Bone Joint Surg Br* 1980;65:51–4.

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