

Case Reports

Fixed Dystonia of the Left Hand in a Violinist: a Rare Functional Disorder

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Abstract

Background: Fixed dystonia leads to an immobile abnormal posturing of the affected limb. There is an ongoing debate whether this condition is psychogenic in origin.

Case report: We present a 21-year-old violinist with fixed dystonia after an acute overuse injury with a transient cyanosis but no signs for psychological trauma. After Incobotulinumtoxin injection, symptoms subsided within 8 hours.

Discussion: Our case corroborates the notion that fixed dystonias after minor injuries are functional disorders. It underlines the necessity of a biopsychosocial approach to functional disorders, considering the possibility of an overlay between organic and non-organic disorders.

Keywords: Fixed dystonia, musician, psychogenic, task specific, hyperkinetic movements

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Introduction

Case report

Musician's dystonia (MD) is a task-specific movement disorder that leads to involuntary cramping with an abnormal posturing of the affected limb when playing an instrument.¹ Fixed dystonia (FD) is a rare manifestation of dystonia that leads to a permanent and immobile abnormal posturing of the affected limb. Its pathophysiology is not well understood; however, it is considered a typical presentation of a functional movement disorder, and risk factors and clinical features have been described.² Here we present a female violinist aged 21 years who suffered from a FD of the left fourth and fifth fingers, which occurred after an injury and resolved within hours of botulinum toxin. This case is interesting for two reasons: first, the course of the disease with onset after a minor injury and a response to botulinum toxin treatment that is incongruous with treatment response in organic dystonia demonstrates that FD is very likely a functional disorder. Secondly, FD is extremely rare in musicians and to our knowledge has not been reported so far.

The patient developed symptoms at the age of 20 years. After playing an Irish traditional music concert that lasted for 5 hours she perceived an involuntary cramping and adduction of the fifth finger of the left hand with cyanosis of the hand. She consequently developed a FD with the fifth finger overlapping the fourth finger, a fixed flexion in the proximal interphalangeal joint (PIP), and extension in the metacarpophalangeal joint (MCP) of both fingers (Figure 1). During the weeks before the concert she had intensely practiced up to 8 hours per day. However, her usual amount of practice was also high, ranging from 4 to 6 hours per day. She started playing the violin at the age of 6 years and has since regularly played in competitions and concerts. She also plays the guitar and the flute.

Medical history revealed two previous episodes of incoordination of the same fingers, which however never turned into a FD. The first episode was in 2008 during a music competition for which she had practiced 6 hours per day and developed a pain syndrome. However,



Figure 1. Posture of the Left Hand on First Presentation of the Patient. When asked to stretch her fingers, the fixed adduction of the fifth finger and the overlapping of the fourth finger above the fifth become visible. There are no difficulties extending digits 2 and 3.



Video 1. The Patient at Her Second Presentation. Violin playing is possible. Still there is a dystonic cramping of digit 5 that reappeared 3 weeks before.

at that time a complete remission occurred under analgesic treatment after 2 weeks. The second episode was in October 2010, again during a time of increased violin playing of up to 10 hours per day, as a result of which she again developed a pain syndrome. Complete remission then occurred after 5 weeks under anesthetic treatment. Family history with regard to neurological disorders was negative.

On neurological examination the hypertonus of the flexor muscles of digits 4 and 5 of the left hand became apparent, leading to the fixed posturing. There was no giving-way of muscle tone upon distraction.³ Violin playing was not possible. There were no sensory deficits, no paresthesia/hypesthesia, and no hyperalgesia/allodynia. Reflexes were normal. There were no signs of edema or abnormal skin blood flow. No Kayser–Fleischer ring was present. Previous history of neuroleptic medication was negative, ruling out a tardive dystonia. Spinal and cerebral magnetic resonance imaging (MRI) scans (T1, T2 with and without contrast) were normal. Electrophysiology (nerve conduction, electromyogram, F-wave) was normal. Blood results including coeruloplasmin, liver profile, vitamin B12, folate, and Lyme serology did not reveal any pathology. There were no previous somatizations in the past, no evidence of a depressive episode or of depression in the past, and no evidence of previous psychological trauma or a secondary benefit.

No improvement was noticed after splinting of the hand for several days, physiotherapy (several times), and acupuncture (twice). L-Dopa+carbidopa (100 mg+25 mg) for 6 weeks, diazepam (10 mg) for 1 week, or local anesthetics (once) did not result in improvement either. Pramipexol (0.088 mg) was not tolerated. We injected 15 MU of Incobotulinumtoxin into the superficial muscle of the fourth and fifth fingers of the left hand as well as 10 MU to the third palmar interosseus muscle. The patient reported a tremendous improvement within 8 hours of injection, which started with a paresthesia (pins and needles) of digits 4 and 5 and led to an almost complete remission within the first days. The effect wore off over the course of about 8–10

weeks, leading to a task-specific dystonic cramping of digit 5 while at the instrument but leaving digit 4 almost normal (Video 1). FD did not reappear.

Discussion

We present the rare case of FD of the left hand in a violinist with the typical manifestation of FD in the fourth and fifth digit and the absence of a sensory trick.² Yet FD is a rare and unusual manifestation of musician's dystonia (MD). Most often MD is task specific dystonia (TSD) and occurs only in the context of playing the instrument. Its typical feature is an involuntary flexion and less often an extension of the affected fingers.^{4,5} In wind instrument players the embouchure may be affected, resulting in involuntary movements of the lips, the tongue, or the jaw.^{4–6} Age of symptom onset is usually in the mid thirties^{7,8} and thus higher than in our patient. In MD, however, the most frequent combination of two affected fingers is digits 4 and 5,⁵ as seen in our patient. One risk factor of MD that has been described is pain due to overuse in up to 9% of patients.^{9,10} The same risk factor is known in FD,^{11–13} where the incidence of pain is considerably higher and occurs in up to 68% of patients.² This is in accordance with our patient who had practiced intensely and for up to 8 hours per day for several weeks prior to symptom onset and consequently developed an acute overuse syndrome. She had had two prior episodes of incoordination 3 years and 1 year ago, which all appeared under similar circumstances of long, intense practice and playing sessions of up to 10 hours per day with a subsequent pain syndrome. However, symptoms always ceased under medication and a break from violin playing. In the context of pain syndromes, the relationship between chronic regional pain syndrome (CRPS) and FD has been repeatedly described.^{14–19} Up to 25% of CRPS patients develop FD¹² and 91% of movement disorders in CRPS are dystonia, of which 75% have FD.¹⁶ It is thus noteworthy that the third episode leading to the FD was

accompanied by transient cyanosis of the affected hand, one main feature of CRPS.²⁰ At clinical examination the criteria for a CRPS were not fulfilled;²⁰ however, this finding is in accordance with reports by Schrag et al,² who found features of CRPS in 44% of patients with FD but only 20% fulfilled the criteria for CRPS.

The most striking feature was the patient's report on an improvement within 8 hours of botulinum toxin injection with an almost complete remission of symptoms within the first days, although anecdotal evidence exists of patients reporting an effect of botulinum toxin within hours. This tremendous improvement is in contrast with reports of a bad outcome of FD accompanied by CRPS features.²¹ This of course raises the question of a possible psychogenic origin of the dystonia. Schrag et al² reported that 36% of patients with FD had a clinically established psychogenic dystonia. Furthermore, the ratio of somatization or conversion disorders was significantly higher in FD than in a control group with non-FD.² Likewise a study reporting on 28 patients with psychogenic movement disorders found that dystonia was the second most common manifestation.²² In one study by Lang²³ of 18 patients with psychogenic dystonia, 61% had had a peripheral trauma in the past, and in a recent paper it was concluded that FD after peripheral trauma is a psychogenic disorder.²⁴ However, another study did not find convincing evidence for a psychogenic genesis of CRPS-related dystonia.²⁵ Finally, psychogenic or functional dystonia is more common in young women after a minor trauma.³ However, there was no previous history for somatization. Nor did she display any signs for a depression, which besides anxiety is the only psychiatric disorder found to be significantly higher in psychogenic movement disorders (PMDs).²⁶ There were no signs of previous psychological trauma, such as abuse or neglect, which from a number of life stressors are almost the only ones significantly more often present in PMDs.²⁶ No secondary benefit was evident. There was no evidence of a factitious disorder or malingering. However, the almost complete remission of symptoms contrasts with reports of a poor prognosis of FD or psychogenic movement disorders²⁴ with more than 75% of patients that do not improve, one-third who continue to deteriorate, and only 6% who have a marked improvement or remission.²¹

Our case demonstrates the rare manifestation of a fixed dystonia in a violin player after acute overuse syndrome. For the following reasons we consider it a functional disorder: first the rapid response to botulinum toxin is incongruous with the treatment effect in organic dystonia; secondly, the rapid onset after a peripheral injury is a common feature in functional movement disorders.^{22,27} Furthermore, complete remissions with sudden recurrences have been reported.³ Finally, functional dystonia occurs most often in young women after an injury.³

The absence of obvious psychogenic signs, psychological trauma, psychiatric disorders, or somatization from our point of view does not contradict the diagnosis because it has been shown that these do not have to be necessarily present in functional movement disorders.^{3,26,28} Our case underlines the necessity of considering a biopsychosocial approach to functional disorders^{3,26,28} that include environmental factors such as, for example, trauma and take into account the

possibility of an overlay between organic and non-organic disorders due to the common occurrence of injury and functional symptoms.^{3,28}

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