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Tremor and Other Hyperkinetic Movements

Case Reports

Characterization and Treatment of Unilateral Facial Muscle Spasm in Linear Scleroderma: A Case Report

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Abstract

Background: Linear scleroderma has been associated with muscle spasms ipsilateral to skin lesions. Typically, spasms are located in trigeminal innervated muscles, leading to hemimasticatory spasm (HMS).

Case Report: We report a case of linear scleroderma associated with spasm of muscles innervated not only by the trigeminal but also by the facial nerve.

Discussion: We review the patient's successful treatment with incobotulinumtoxinA, a formulation of botulinum toxin that has not been reported for use in this condition.

Keywords: Hemimasticatory spasm, botulinum toxin, incobotulinumtoxinA, hemifacial spasm, dystonia, linear scleroderma

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Introduction

Linear scleroderma is a chronic connective tissue disease characterized by skin lesions in the face and neck. These lesions are characterized as en coup de sabre, vertical, colorless skin indentations that resemble a wound from being struck by a sword.[1,2] These involve the deep and superficial layers of the skin and can impair motion of the underlying joints. Linear scleroderma has been associated with hemimasticatory spasm (HMS) in a number of cases.[3]

HMS is characterized by involuntary muscle spasms of the jaw-closing muscles on one side of the face.[4] One defining electrophysiological characteristic of HMS is the absence of silent periods during times of involuntary spasms.[5] HMS is thought to be due to focal demyelination at the motor root or the motor nucleus of the trigeminal nerve,[6] or from injury to the motor fibers of the trigeminal nerve from deep tissue changes caused by linear scleroderma[7] or dental procedures.[3] It affects the trigeminal innervated muscles, including the temporalis, masseter, and medial and lateral pterygoid muscles.[8]

Hemifacial spasm (HFS) is a far more common condition characterized by spasms of the muscle fibers innervated by the ipsilateral facial nerve.[9] The two types of HFS include idiopathic and secondary HFS. Idiopathic HFS is in some cases thought to be caused by nerve compression at the root exit point whereas secondary HFS can be triggered by any other damage to the facial nerve, including prior Bell's palsy, effects of demyelination in multiple sclerosis, and from conditions known to cause cranial neuropathies such as Lyme disease and sarcoidosis. The average age of onset is 44.[10]

Botulinum toxin A has been successfully used to treat symptoms of dystonia, HFS, and HMS due to a variety of etiologies.[11–13]

Case report

We report a previously healthy Caucasian man diagnosed at age 30 with linear scleroderma. He developed facial spasms about 2 years after the onset of the skin lesions. Both the skin lesions and the muscle spasms occurred only on the left side. The patient was aware of these muscle contractions, which led to emotional distress and social withdrawal.

The muscle spasms at first occurred randomly, and after a few months became nearly constant throughout the waking day; these spasms resulted in teeth chattering and involuntary movements of the jaw up to several times an hour.

The spasms were exacerbated by touch such as lying on the left side of his face. These spasms could be suppressed for a couple of seconds by the patient tightening all his upper and lower facial musculature into a forceful grimace and were partially relieved by chewing, such as the use of gum. The spasms caused considerable discomfort, but were not painful. There was no urge to move or a build-up of inner tension before these involuntary muscle contractions. No sensory symptoms were reported by the patient.

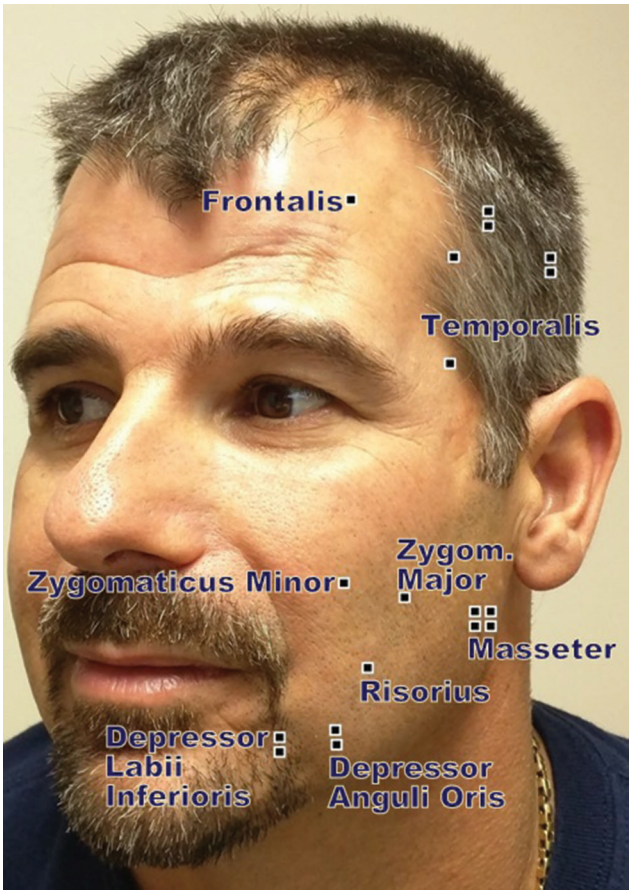

Figure 1. **Typical Injection Sites under EMG Guidance**. IncobotulinumtoxinA. . = 2.5 u/0.05 mL; : = 5 u/0.1 mL; :: = 10 u/0.2 mL.

General examination revealed a band of sclerotic skin over the left brow and along the left corner of the mouth. Muscle spasms presented as rippling movements beneath the skin in the muscles identified clinically as the left temporalis, masseter, frontalis, depressor supercilii, risorius, zygomaticus, depressor labii inferioris, and depressor anguli oris. The left temporalis and masseter muscles were markedly hypertrophic.

Myokymic discharges and dystonic muscle contractions were identified based on the typical audio signature on the needle electromyography of grouped repetitive discharges and sustained tonic discharges. Dystonic contractions were predominantly noted in the masseter and temporalis; myokymic discharges and non-sustained tonic discharges were noted in the frontalis, risorius, zygomaticus, and depressor anguli oris muscles.

Thus far, the patient has received 22 injection sessions of incobotulinumtoxinA (Xeomin, Merz) over the past 6 years. His initial dose was 30 units and it has been increased over time to 45 units (Figure 1). A seven-point semiquantitative outcome scale was used before each of the 22 injection sessions (0 = no disability to 6 = completely incapacitated) (Table 1). Scoring was based on patient report and the examiner's assessment. This analysis is performed routinely in the clinic before all botulinum toxin injections, in all patients, for any indication.

- The patient scored 5 (severe disability or functional impairment) before the initial injection.
- At his best level of function after receiving botulinum toxin injections, the patient averaged a score of 0.14 across 22 injections.

Table 1. **Outcome Scale Based on Patient Report and Examiner's Assessment Performed at Each Botulinum Injection Session**

Current (pre-injection) disability
0 No disability or functional impairment
1 Mild disability or functional impairment
2 Mild–moderate disability or functional impairment
3 Moderate disability or functional impairment
4 Moderate–severe disability or functional impairment
5 Severe disability or functional impairment
6 Completely incapacitated
Disability at patient's best following last botulinum toxin injection session
0 No disability or functional impairment
1 Mild disability or functional impairment
2 Mild–moderate disability or functional impairment
3 Moderate disability or functional impairment
4 Moderate–severe disability or functional impairment
5 Severe disability or functional impairment
6 Completely incapacitated

- The duration of effect of each injection was approximately 10.9 weeks.
- There was an average of 15.1 weeks between each injection.
- At the time of injection, the effects of botulinum toxin injections had partially worn off and the patient averaged a score of 3.73 across the 22 injections.

Since the start of the injections, the patient has experienced marked relief from the involuntary muscle spasms and hypertrophy.

Discussion

There have been no reports on the effective use of incobotulinumtoxinA in HMS. IncobotulinumtoxinA is a low molecular weight formulation of botulinum toxin type A free of non-toxin complexing proteins.

As opposed to classically defined HMS, non-trigeminal innervated muscles have been identified in our patient, and have also been injected with botulinum toxin with good results. There have only been a few cases of HMS described in the literature, and only one case, to our knowledge, in which non-trigeminal innervated muscles are involved.[14]

Our report suggests that the effects of linear scleroderma can extend to other non-trigeminal innervated muscle groups, which would support the proposed pathophysiologic mechanism of localized injury to the motor fibers from the deep tissue changes caused by linear scleroderma. We show that botulinum toxin therapy with incobotulinumtoxinA can be used safely and effectively over several years.

References

1. Hawk A, English JC 3rd. Localized and systemic scleroderma. *Semin Cutan Med Surg* 2001;20:27–37. doi: 10.1053/sder.2001.23093

2. Jun JH, Kim HY, Jung HJ, Lee WJ, Lee SJ, Kim DW, et al. Parry-Romberg syndrome with en coup de sabre. *Ann Dermatol* 2011;23:342–347. doi: 10.5021/ad.2011.23.3.342

3. Kaufman MD. Masticatory spasm in facial hemiatrophy. *Ann Neurol* 1980; 7:585–587. doi: 10.1002/ana.410070614

4. Chon K, Lee J, Koh E, Choi H. Hemimasticatory spasm treated with microvascular decompression of the trigeminal nerve. *Acta Neurochirurgica* 2012; 154:1635–1639. doi: 10.1007/s00701-012-1360-y

5. Ongerboer De Visser BW, Cruccu G, Manfredi M, Koelman JH. Effects of brainstem lesions on the masseter inhibitory reflex: functional mechanisms of reflex pathways. *Brain* 1990;113:781–792. doi: 10.1093/brain/113.3.781

6. Cruccu G, Inghilleri M, Berardelli A, Pauletti G, Casali C, Coratti P, et al. Pathophysiology of hemimasticatory spasm. *J Neurol Neurosurg Psychiatry* 1994;57: 43–50. doi: 10.1136/jnnp.57.1.43

7. Kim HJ, Jeon BS, Lee K. Hemimasticatory spasm associated with localized scleroderma and facial hemiatrophy. *Arch Neurol* 2000;57:576–580. doi: 10.1001/archneur.57.4.576

8. Christie C, Rodríguez-Quiroga SA, Arakaki T, Rey RD, Garretto NS. Hemimasticatory spasm: report of a case and review of the literature. *Tremor Other Hyperkinet Mov* 2014;4. doi: 10.7916/D8QF8QWD

9. Yaltho TC, Jankovic J. The many faces of hemifacial spasm: differential diagnosis of unilateral facial spasms. *Mov Disord* 2011;26:1582–1592. doi: 10.1002/mds.23692

10. Lu AY, Yeung JT, Gerrard JL, Michaelides EM, Sekula RF, Bulsara KR. Hemifacial spasm and neurovascular compression. *Sci World J* 2014;2014: 349319. doi: 10.1155/2014/349319

11. Wickwar S, McBain H, Newman SP, Hirani SP, Hurt C, Dunlop N, et al. Effectiveness and cost-effectiveness of a patient-initiated botulinum toxin treatment model for blepharospasm and hemifacial spasm compared to standard care: study protocol for a randomised controlled trial. *Trials* 2016; 17:129. doi: 10.1186/s13063-016-1263-y

12. Kim JH, Han SW, Kim YJ, Kim J, Oh MS, Ma HI, et al. A case of painful hemimasticatory spasm with masseter muscle hypertrophy responsive to botulinum toxin. *J Mov Disord* 2009;2:95–97. doi: 10.14802/jmd.09026

13. Mir P, Gilio F, Edwards M, Inghilleri M, Bhatia KP, Rothwell JC, et al. Alteration of central motor excitability in a patient with hemimasticatory spasm after treatment with botulinum toxin injections. *Mov Disord* 2006;21:73–78. doi: 10.1002/mds.20653

14. Cañas CA, Orozco JL, Paredes AC, Bonilla-Abadía F. Successful treatment of hemifacial myokymia and dystonia associated to linear scleroderma "en coup de sabre" with repeated botox injections. *Case Rep Med* 2012;2012: 691314. doi: 10.1155/2012/691314