Hemimasticatory Spasm: Report of a Case and Review of the Literature

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Abstract

Background: Hemimasticatory spasm is a very rare movement disorder characterized by unilateral, involuntary, paroxysmal contractions of the jaw-closing muscles, causing clinically brief twitches and/or spasms.

Case Report: A 62-year-old female consulted us with a 30-year history of unusual involuntary twitches in the preauricular region and spasms that hampered jaw opening. During these spasms, she could not open her mouth. On physical examination, we also observed hypertrophy of the masseter and temporalis muscles, which can be features of hemimasticatory spasm. She was treated with botulinum toxin type A, with excellent response. Here, we present her case and review the literature.

Discussion: Hemimasticatory spasm is a rare movement disorder. Given the excellent response to botulinum toxin type A treatment, it should be considered within the spectrum of facial spasms.

Keywords: Hemimasticatory spasm, botulinum toxin, jaw-closing spasm

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Conflict of Interest: The authors report no conflict of interest.
| Authors                          | Age at Onset | Sex | Involved Muscles                      | Mechanism or Special Clinical Features | Response to Botulinum Toxin | Surgical Treatment |
|---------------------------------|--------------|-----|--------------------------------------|----------------------------------------|----------------------------|-------------------|
| Kaufman, 1980                   | 25           | F   | Left masseter                        | —                                      | NA                         |                   |
| Lapresle, 1982                  | 15           | F   | Right masseter                       | Linear scleroderma with right FHA      | NA                         | —                 |
| Thompson and Carroll, 1983      | 57           | F   | Left masseter and temporalis         | Idiopathic                             | NA                         | Cryosurgical lesion |
| Thompson, et al., 1986          | 31           | F   | Right masseter                       | Morphea with right FHA                 | NA                         | Myotomy           |
| Parisi, et al., 1987            | 38           | F   | Right masseter                       | Linear scleroderma with right FHA      | NA                         |                   |
| Yoshii and Alba, 1989           | 44           | M   | Left masseter and both (medial and lateral) pterygoids | Idiopathic                             | NA                         |                   |
| Auger, et al., 1992             | 20           | F   | Right masseter and temporalis        | Idiopathic                             | Yes                        | Transient response to trigeminal rootlets section |
|                                 | 17           | F   | Right medial pterygoid               | Idiopathic                             | NA                         |                   |
|                                 | 20           | F   | Right masseter and temporalis        | Idiopathic                             | NA                         |                   |
| Cruccu, et al., 1994            | 18           | M   | Left temporalis                      | Left FHA                               | NA                         |                   |
|                                 | 44           | F   | Right masseter and temporalis        | Morphea                                | Yes                        |                   |
|                                 | 44           | M   | Right masseter                       | FHA                                    | Yes                        |                   |
| Ebersbach, et al., 1995         | 26           | M   | Left masseter and temporalis         | Left FHA                               | Yes                        |                   |
|                                 | 26           | F   | Right masseter and temporalis        | Local scleroderma with FHA             | Yes                        |                   |
|                                 | 34           | F   | Right masseter                       | Local scleroderma with FHA             | Yes                        |                   |
|                                 | 47           | F   | Left masseter                        | Idiopathic                             | Yes                        |                   |
|                                 | 44           | F   | Right masseter and temporalis        | Idiopathic                             | Yes                        |                   |
| Wang, et al., 2004              | 38           | F   | Left masseter                        | NA                                     | NA                         |                   |
|                                 | 12           | M   | Right masseter and temporalis        | Right linear scleroderma               | NA                         |                   |
|                                 | 33           | M   | Right temporal                       | NA                                     | NA                         |                   |
Table 1. Continued

| Authors                      | Age at Onset | Sex | Involved Muscles          | Mechanism or Special Clinical Features | Response to Botulinum Toxin | Surgical Treatment |
|------------------------------|--------------|-----|---------------------------|----------------------------------------|-----------------------------|--------------------|
| 21                           | 42           | F   | Left masseter and temporalis | NA                                     | NA                          | —                  |
| Cersosimo, et al. 2003       | 29           | F   | Right masseter and temporalis | Severe worsening during pregnancy      | Yes                         | —                  |
| Mir, et al. 2006             | 26           | M   | Left masseter and temporalis | Idiopathic                             | Yes                         | —                  |
| Gunduz, et al. 2007          | 62           | F   | Right masseter and temporalis | Right pontine and cerebellar hemisphere infarction | Yes                         | —                  |
| Jiménez-Jiménez, et al. 2007 | 40           | M   | Right masseter and temporalis | Biopercular infarct with previous Foix–Marie–Chavany syndrome | Yes                         | —                  |
| Kumar, et al. 2008           | 49           | F   | Left masseter, temporalis and lateral pterygoid | Left morphea                           | Yes                         | —                  |
| Yalto and Jankovic 2011      | 63           | F   | Left masseter             | Idiopathic                             | Yes                         | —                  |
| Gopalakrishnan, et al. 2011  | 56           | F   | Left masseter and temporalis | Cerebellopontine angle hematoma        | Spontaneous remission       | —                  |
| Sinha, et al. 2011           | 38           | M   | Right masseter and temporalis | Idiopathic                             | —                           | Debulking and stripping masseter muscle |
| Chon, et al. 2012            | 40           | M   | Right masseter and temporalis | Idiopathic                             | Yes                         | MVD                |
| Wang, et al. 2013            | 50           | F   | Left masseter             | NA                                     | NA                          | MVD                |
| 32                           | 42           | F   | Right masseter and temporalis | NA                                     | NA                          | MVD                |
| 33                           | 38           | M   | Right masseter             | NA                                     | NA                          | MVD                |
| 34                           | 48           | F   | Right masseter             | NA                                     | NA                          | MVD                |
| 35                           | 57           | F   | Left masseter and temporalis | NA                                     | NA                          | MVD                |
| 36                           | 53           | F   | Right masseter and temporalis | NA                                     | NA                          | MVD                |
| 37                           | 32           | F   | Right masseter             | Idiopathic                             | Yes                         | —                  |

FHA, Facial Hemiatrophy; F, Female; M, Male; MVD, Microvascular Decompression; NA, Not Available.
On physical examination, hypertrophy of the masseter and temporalis muscles was noted. Dental treatments were required in order to repair several broken teeth.

She has no other medical conditions or family history; she does not take any medications and has no laboratory evidence for connective tissue disease or thyroid dysfunction.

At present, computerized tomography scan of the brain, brain magnetic resonance imaging, and electroencephalography are normal. Electromyography (EMG) of the right masseter and temporalis muscles revealed spontaneous activity consisting of repetitive, spontaneous bursts of motor unit discharges, ranging from 100 to 200 Hz (Figure 1).

Over the last years, she has been treated with injections of botulinum toxin type A, every 3–4 months, with 60 U in the right masseter muscle and 40 U in the right temporalis muscle, with an excellent response. To date, this treatment remains beneficial.

**Discussion**

HMS is characterized by involuntary movements, consisting of brief twitches and/or spasms, resembling cramps. It is considered a disorder of the motor branch of the trigeminal nerve, and is characterized by unilateral, involuntary, paroxysmal, sometimes painful, violent, and prolonged contractions of the jaw-closing muscles.3,4

Typically, HMS involves the masseter and the temporalis muscles, with the medial pterygoid muscle also rarely being involved. There is usually no involvement of the jaw-opening muscles, but there are at least two cases describing involvement of the lateral pterygoid (Table 1), one of them with associated lateral deviation of the jaw.5 There are no reports of bilateral involvement. HMS more commonly presents in females in the third and fourth decade, as observed in our patient.4

The most frequent triggers that precipitate spasms are talking, laughing, or chewing; these triggers are always voluntary movements rather than sensory stimuli, as in trigeminal neuralgia. Brief spasms are generally painless; prolonged spasms can be painful, as occurs with cramps. Severe or violent spasms can result in temporomandibular joint dislocation, and some patients, such as ours, may even bite their tongue or break teeth.6

The neurological examination should be normal in HMS, except for the spasm, the hypertrophy of the involved muscles or the atrophy of the subcutaneous tissue that may occur in cases associated with localized scleroderma.7 Facial sensation is always spared and no other cranial nerves should be compromised.

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**Video 1. Hemimasticatory Spasm in a 62-year-old Female.** The patient presents involuntary contraction of the right temporal and masseter muscles.

**Figure 1. Electromyographic recording.** Simultaneous electromyography recording (concentric needle electrodes) from right masseter and temporalis muscles shows continuous bursts of activity during the prolonged spasms.
The pathophysiologic mechanisms that produce HMS are not entirely clear. There is an impaired inhibition of the muscle contraction that can be evidenced electrically by loss of the silent period, which is almost unique to HMS, and so can be a very useful aid for differential diagnosis.\(^5\,^6\) The characteristic EMG findings of HMS include irregular bursts of motor unit potentials (MUPs) that correlate with the involuntary masseter spasms.

MUPs are often morphologically normal but with very high frequency. Cruccu et al.\(^4\) noticed a delay in the conduction speed of the motor branch of the trigeminal nerve, localized at the infratemporal fossa between the lateral pterygoid and skull surface. This could explain a focal demyelination of the trigeminal motor fibers in these cases, as well as the hemifacial atrophy seen in almost 70% of cases.

In HMS, unlike unilateral jaw closing oro-mandibular dystonia, there is no agonist/antagonist muscle co-contraction during the voluntary movement of jaw opening.\(^7\) Furthermore, electrophysiological studies have demonstrated that the masseter inhibitory reflex and the silent period were absent during periods of spasm in the affected side, independent of the stimulated trigeminal nerve. The complete absence of the silent period in one or more muscles of one side of the face is an almost exclusive feature of HMS.\(^8\)

The fact that in almost all informed cases the muscles affected were the masseter and temporal, sometimes the medial pterygoid, but only on two occasions the lateral pterygoid, suggests that the site generator of ectopic impulses should be at the distal fibers of the trigeminal nerve.\(^4\) This is also supported by previous reports showing relief by microvascular decompression of the trigeminal nerve.\(^9\)

The hypertrophy of jaw-closing muscles, as in our patient, suggests that the generator of ectopic impulse may be at the motor root of the trigeminal nerve or at its motor nucleus, as can be seen in hemifacial spasm.\(^6\,^8\)

Botulinum toxin type A injection is the most effective available treatment.\(^10\,^11\)

In summary, HMS is a rare movement disorder. Given the excellent response to botulinum toxin type A treatment, it should be considered within the spectrum of focal spasms.

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