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Tremor and Other Hyperkinetic Movements

Case Reports

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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Introduction

Unlike hemifacial spasm, hemimasticatory spasm (HMS) is a very rare disorder. Since the original description by Gowers in 1897,[1] there have been very few cases described in the worldwide literature. In a recent study in a Movement Disorders Clinic, conducted between 2000 and 2010, there was only one HMS patient among 215 patients with hemifacial spasm.[2] To our knowledge, only 36 HMS cases have been reported in the literature between 1980 and 2013 (Table 1).

The cause of HMS is not fully known, but it is considered the result of a problem in the motor branch of the trigeminal nerve, and it is characterized by unilateral, involuntary, paroxysmal contractions of the jaw-closing muscles, causing clinically brief twitches and/or spasms.

We describe a patient with a severe form of HMS with good response to botulinum toxin type A treatment.

Aside from reporting a rare entity and reviewing the literature, we draw attention to the long natural history of the condition before the patient was correctly diagnosed and treated. This highlights two issues: firstly, the delay in diagnosis due to the rarity of the condition, and, secondly, the confirmation that spasms remained at the same topography for many years, during which time only the frequency and intensity of the spasms increased.

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Case Report

A 62-year-old female presented with a 30-year history of strange sensations in the right preauricular region, and involuntary contractions of the jaw, causing sporadic biting injuries to her buccal mucosa. Initially, laughing, talking, or eating were the precipitant factors for these contractions.

The symptoms progressed gradually so that at present she has severe daily contractions, with up to 30 episodes per day. The episodes consist of spontaneous appearance of twitches in the preauricular region, or spasms that hamper jaw opening for as long as 20 seconds. Temporomandibular pain is only present during prolonged spasms or contractions (Video 1).

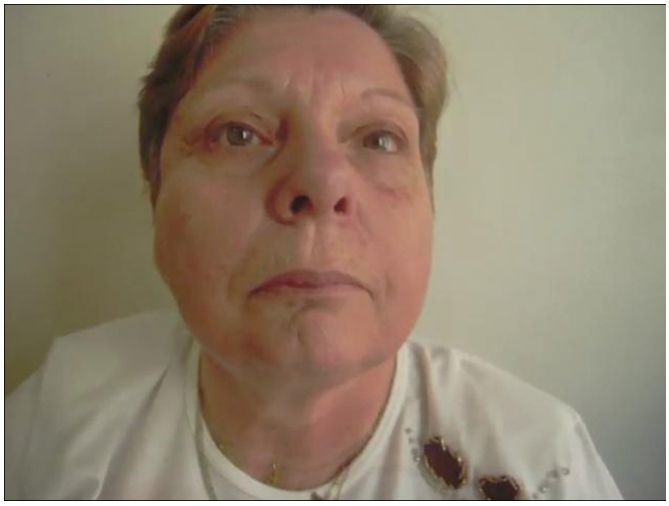
Table 1. Summary of Reported Cases of Hemimasticatory Spam in the Literature

	Authors	Age at Onset	Sex	Involved Muscles	Mechanism or Special Clinical Features	Response to Botulinum Toxin	Surgical Treatment
1	Kaufman, 1980[12]	25	F	Left masseter	—	NA	—
2	Lapresle 1982[13]	15	F	Right masseter	Linear scleroderma with right FHA	NA	—
3	Thompson and Carroll 1983[14]	57	F	Left masseter and temporalis	Idiopathic	NA	Cryosurgical lesion
4	Thompson, et al. 1986[6]	31	F	Right masseter	Morphea with right FHA	NA	Myotomy
5	Parisi, et al. 1987[15]	38	F	Right masseter	Linear scleroderma with right FHA	NA	—
6	Yoshii and Alba 1989[16]	44	M	Left masseter and both (medial and lateral) pterygoids	Idiopathic	NA	—
7	Auger, et al. 1992[3]	20	F	Right masseter and temporalis	Idiopathic	Yes	Transient response to trigeminal rootlets section
8		17	F	Right medial pterygoid	Idiopathic	NA	—
9		20	F	Right masseter and temporalis	Idiopathic	NA	—
10	Cruccu, et al. 1994[4]	18	M	Left temporalis	Left FHA	NA	—
11		44	F	Right masseter and temporalis	Morphea	Yes	—
12	Kim, et al. 1994[17]	44	M	Right masseter	FHA	Yes	—
13	Ebersbach, et al. 1995[18]	26	M	Left masseter and temporalis	Left FHA	Yes	—
14		26	F	Right masseter and temporalis	Local scleroderma with FHA	Yes	—
15	Kim, et al. 2000[7]	34	F	Right masseter	Local scleroderma with FHA	Yes	—
16	Esteban, et al. 2002[19]	47	F	Left masseter	Idiopathic	Yes	—
17	Teive, et al. 2002[11]	44	F	Right masseter and temporalis	Idiopathic	Yes	—
18	Wang, et al. 2004[20]	38	F	Left masseter	NA	NA	—
19		12	M	Right masseter and temporalis	Right linear scleroderma	NA	—
20		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	—
22	Cersosimo, et al. 2003[10]	29	F	Right masseter and temporalis	Severe worsening during pregnancy	Yes	—
23	Mir, et al. 2006[21]	26	M	Left masseter and temporalis	Idiopathic	Yes	—
24	Gunduz, et al. 2007[22]	62	F	Right masseter and temporalis	Right pontine and cerebellar hemisphere infarction	Yes	—
25	Jiménez-Jiménez, et al. 2007[23]	40	M	Right masseter and temporalis	Biopercular infarct with previous Foix–Marie–Chavany syndrome	Yes	—
26	Kumar, et al. 2008[5]	49	F	Left masseter, temporalis and lateral pterygoid	Left morphea	Yes	—
27	Yaltho and Jankovic 2011[2]	63	F	Left masseter	Idiopathic	Yes	—
28	Gopalakrishnan, et al. 2011[24]	56	F	Left masseter and temporalis	Cerebellopontine angle hematoma	Spontaneous remission	—
29	Sinha, et al. 2011[25]	38	M	Right masseter and temporalis	Idiopathic	—	Debulking and stripping masseter muscle
30	Chon, et al. 2012[26]	40	M	Right masseter and temporalis	Idiopathic	Yes	MVD
31	Wang, et al. 2013[9]	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	This case Christie, et al. 2014	32	F	Right masseter	Idiopathic	Yes	—

FHA, Facial Hemiatrophy; F, Female; M, Male; MVD, Microvascular Decompression; NA, Not Available.



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

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.[4] Brief spasms are generally painless; prolonged spasms can be painful, as occurs with cramps. Severe or violent spasms can result in temporo-mandibular 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.

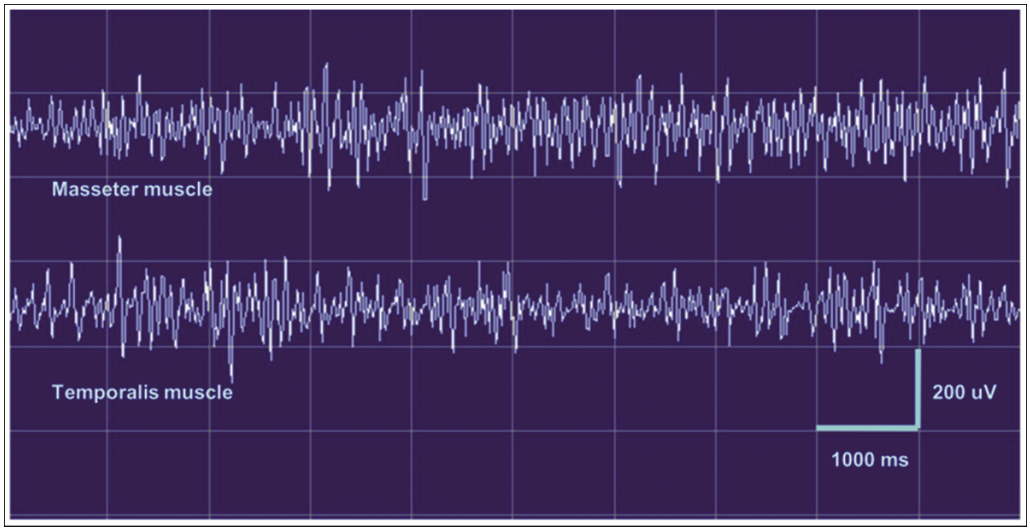


Figure 1. **Electromyographic recording.** Simultaneous electromyography recording (concentric needle electrodes) from right masseter and temporalis muscles shows continuous bursts of activity during the prolonged spasms.

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.[4,6] 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.[3] 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 facial spasms.

References

1. Gowers WR. *A manual of diseases of the nervous system.* 1897, 2nd edition, Philadelphia: Blakiston. Vol 2. p 221–224.

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

3. Auger RG, Litchy WJ, Cascino TL, Ahlskog E. Hemimasticatory spasm: Clinical and electrophysiologic observations. *Neurology* 1992;42:2263–2266, doi: http://dx.doi.org/10.1212/WNL.42.12.2263

4. Cruccu G, Inghilleri M, Berardelli A, et al. Pathophysiology of hemimasticatory spasm. *J Neurol Neurosurg Psychiatry* 1994;57:43–50, doi: http://dx.doi.org/10.1136/jnnp.57.1.43.

5. Kumar N, Krueger BR, Ahlskog JE. Hemimasticatory spasm with lateral jaw deviations. *Mov Disord* 2008;23:2265–2266, doi: http://dx.doi.org/10.1002/mds.22304.

6. Thompson PD, Obeso JA, Delgado G, Gallego J, Marsden CD. Focal dystonia of the jaw and the differential diagnosis of unilateral jaw and masticatory spasm. *J Neurol Neurosurg Psychiatry* 1986;49:651–656, doi: http://dx.doi.org/10.1136/jnnp.49.6.651.

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

8. Micheli F, Luquin-Piudo MR. *Movimientos anormales: Clínica y terapéutica.* 1ra edición. 2012: Editorial: Panamericana. Chapter 36: "Espasmo Hemifacial y Espasmo Hemimasticatorio." P. 549–559.

9. Wang YN, Dou NN, Zhou QM, et al. Treatment of hemimasticatory spasm with microvascular decompression. *J Craniofac Surg* 2013;24:1753–1755, doi: http://dx.doi.org/10.1097/SCS.0b013e318295025a.

10. Cersosimo MG, Bertoti A, Roca CU, Micheli F. Botulinum toxin in a case of hemimasticatory spasm with severe worsening during pregnancy. *Clin Neuropharmacol* 2004;27:6–8, doi: http://dx.doi.org/10.1097/00002826-200401000-00004.

11. Teive HA, Piovesan EJ, Germiniani FMB, et al. Hemimasticatory spasm treated with botulinum toxin: Case report. *Arq Neuropsiquiatr* 2002;60:288–289, doi: http://dx.doi.org/10.1590/S0004-282X2002000200020.

12. Kaufman MD. Masticatory spasm in facial hemiatrophy. *Ann Neurol* 1980;7:585–587, doi: http://dx.doi.org/10.1002/ana.410070614.

13. Lapresle J, Desi M. Sclerodermie avec hemiatrophie faciale progresive et atrophie croisee de 1 hemicorps. *Rev Neurol (Paris)* 1982;138:815–822.

14. Thompson PD, Carroll WM. Hemimasticatory and hemifacial spasm: A common pathophysiology? *Clin Exp Neurol* 1983;19:110–119.

15. Parisi L, Valente G, Dell'Anna C, Mariorenzi R, Amabile G. A case of facial hemiatrophy associated with linear scleroderma and homolateral masseter spasm. *Ital J Neurol Sci* 1987;8:63–65, doi: http://dx.doi.org/10.1007/BF02361438.

16. Yoshii K, Seki Y, Aiba T. A case of unilateral masticatory spasm without hemifacial atrophy. *No To Shinkei* 1989;57:43–50.

17. Kim YH, Lee KS, Na JH, Kim BS, Ko YJ. A case of hemimasticatory spasm. *J Korean Neurol Assoc* 1994;12:175–178.

18. Ebersbach G, Kabus C, Schelosky L, Terstegge L, Poewe W. Hemimasticatory spasm in hemifacial atrophy: Diagnostic and therapeutic aspects in two patients. *Mov Disord* 1995;10:504–507, doi: http://dx.doi.org/10.1002/mds.870100417.

19. Esteban A, Traba A, Prieto J, Grandas F. Long term follow up of a hemimasticatory spasm. *Acta Neurol Scand* 2002;105:67–72, doi: http://dx.doi.org/10.1034/j.1600-0404.2002.00119.x.

20. Wang YW, Ma X, Zhang ZK, Shen D, Su F, Fu K. [Hemimasticatory muscle spasm: An electromyogram analysis]. *Zhonghua Kou Qiang Yi Xue Za Zhi* 2004;39:155–157.

21. Mir P, Gilio F, Edwards M, 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: http://dx.doi.org/10.1002/mds.20653.

22. Gunduz A, Karaali-Savrun F, Uluduz D. Hemimasticatory spasm following pontine infarction. *Mov Disord* 2007;22:1674–1675, doi: http://dx.doi.org/10.1002/mds.21406.

23. Jimenez-Jimenez FJ, Puertas I, Alonso-Navarro H. Hemimasticatory spasm secondary to biopercular syndrome. *Eur Neurol* 2008;59:276–279, doi: http://dx.doi.org/10.1159/000115644.

24. Gopalakrishnan CV, Dhakoji A, Nair S. Hemimasticatory spasm following surgery for vestibular schwannoma. *Mov Disord* 2011;26:2481–2482, doi: http://dx.doi.org/10.1002/mds.23988.

25. Sinha R, Chattopadhyay PK. Hemimasticatory spasm: A case report with a new management strategy. *J Maxillofac Oral Surg* 2011;10:170–172, doi: http://dx.doi.org/10.1007/s12663-010-0162-0.

26. Chon KH, Lee JM, Koh EJ, Choi HY. Hemimasticatory spasm treated with microvascular decompression of the trigeminal nerve. *Acta Neurochir (Wien)* 2012;154:1635–1639, doi: http://dx.doi.org/10.1007/s00701-012-1360-y.