

Hemi masticatory spasm with facial hemi atrophy and localized scleroderma: Report of a case with bilateral involvement

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Abstract

We report a rare case of a 45 year old female with 15 year history of progressive left facial thinning with frequent episodes of involuntary jaw closure and almost continuous rippling movements over her left sided masticatory muscles. There was localized scleroderma, left facial hemi atrophy and left hemi masticatory spasm. Localized scleroderma was proven histopathologically. Electrophysiological studies revealed normal blink reflex on both sides. Her masseter inhibitory reflex was absent bilaterally and surface electromyogram showed spontaneous bursts of high frequency activity over bilateral masseter and left temporalis muscles. The patient responded remarkably with bilateral botulinum toxin injection. This case highlights presence of rare bilateral involvement of HMS especially on EMG and excellent response to botulinum toxin.

Keywords: hemi masticatory spasm (HMS), scleroderma, facial hemi atrophy, bilateral, botulinum toxin

INTRODUCTION

Hemi masticatory spasm (HMS) is a rare movement disorder characterized by unilateral pain and spasm of the masticatory muscles due to dysfunction of motor branches of trigeminal nerve. Proposed pathogenesis is focal demyelination of motor part of trigeminal nerve due to microvascular compression (as in hemifacial spasm) leading to spasm of supplied muscles.¹⁻³ HMS usually affects women in third to fourth decades and there is association with hemi atrophy in two thirds and localized scleroderma in one third of cases.²⁻⁴ We present a case of HMS with evidence of asymmetric bilateral involvement, treated with botulinum toxin.

CASE REPORT

A 45-year-old lady presented to us with a 15-year history of gradually progressive thinning and dark pigmentation of skin over the left side of forehead, reaching up to the vertex. This was associated with gradual and progressive loss of hair over the scalp on left side along with atrophy of left half of face over 10 years (resulting in facial asymmetry). Ten years back she developed episodic difficulty in opening her mouth, lasting 1-2 hours, several times a day. These spasms were painless and triggered by chewing, speaking, tapping on jaw or

voluntary clenching of teeth. Over the last 5 years frequency and duration of spasms increased and would appear spontaneously, making talking and swallowing very difficult. There was no deviation of jaw during this spasm. She had also noticed appearance of rippling movements over the left half of her face, mainly over the temple and jaw, which would become very prominent during the episodes of jaw closure (video 1).

A magnetic resonance imaging study of the brain was normal and skin biopsy revealed changes of localized scleroderma. Surface electromyography (EMG) studies revealed intermittent spontaneous high frequency (100-200 Hz) activity recorded simultaneously from bilateral masseter and left temporalis muscles (Figure 1a). Her blink reflex was normal on both the sides. The masseter-inhibitory reflex revealed loss of inhibition in the form of absent silent period, on testing over both sides (Figure 1b). Since EMG had revealed spontaneous bursts of high frequency activity over bilateral masseter and left temporalis muscles, the patient was treated with injection botulinum toxin A (left temporalis 8 units and masseters right = 20 IU/ left = 30 IU, Botox, Allergan). Marked improvement was noted in the form of relief from episodes of jaw closure from the third day onwards after injection and 90% reduction in the spasm (global subjective

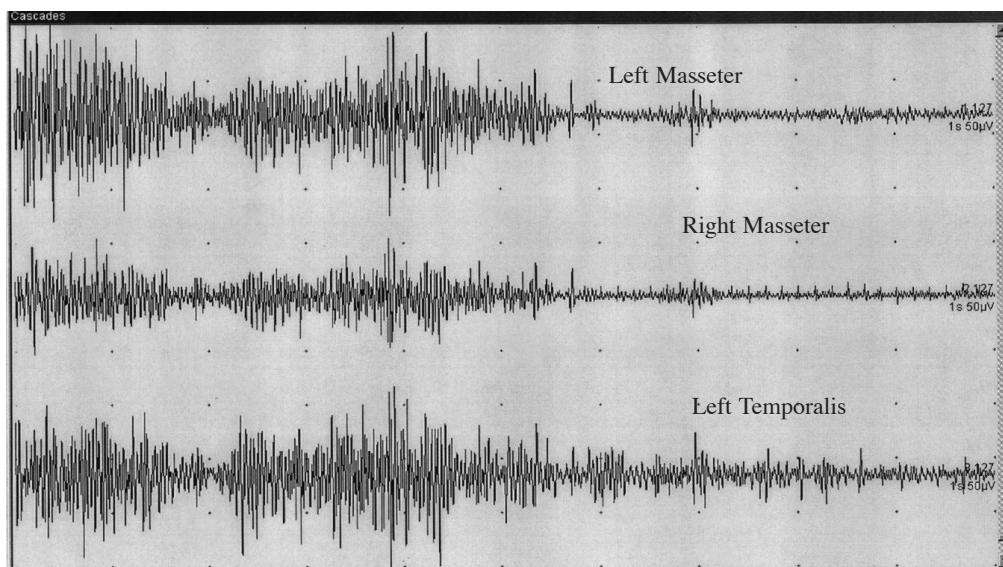


Figure 1a. Surface EMG recording over left masseter, right masseter and left temporalis muscles (from top to bottom) showing simultaneous contraction and relaxation over all muscles, during spasm. Recording has been made with sensitivity of $50\mu\text{V}$ and at a sweep speed of 1 second per division.

assessment scale) over the injected muscles. Effect of the first injection lasted for 3 months, after which repeated injection gave almost 100% improvement in both jaw closure as well as the visible rippling of the muscles.

DISCUSSION

We describe a patient with HMS, facial

hemi atrophy and localized scleroderma, an extremely rare disorder. The chief aim of this report is to focus on the evidence of bilateral involvement demonstrated electro physiologically, as demonstrated by EMG, and excellent response to botulinum toxin injection.

The clinical presentation of all cases described in previous reports is with two types of involuntary movements of muscles of jaw closure (temporalis,

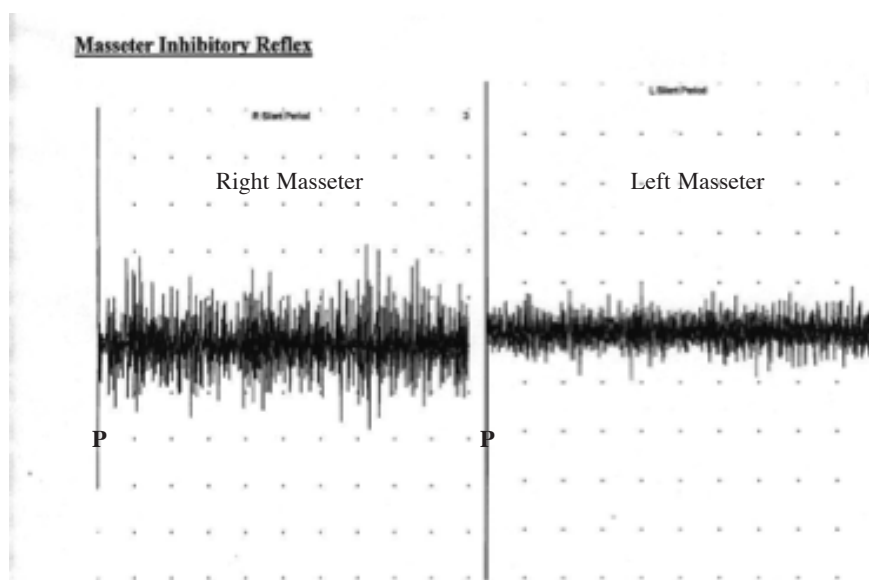


Figure 1b. On testing for the masseter inhibitory reflex, no silent period was recorded on both sides. "P" indicates the point of stimulation.

masseter and occasionally medial pterygoid).¹⁻⁸ One of these movements consists of brief involuntary twitches of these muscles, and the second being sudden onset, short or prolonged painful spasms of these muscles, often precipitated by chewing, talking, yawning or other voluntary movements of mouth and jaw. The involuntary movements of the current patient differ considerably from previously reported cases. She experienced very prolonged but painless spasms, lasting several hours. These were associated with almost continuous rippling movements over her affected temporalis and masseter muscles.

HMS is associated with facial hemi atrophy in 70% and morphea in 40% cases.⁴ Facial hemiatrophy many times involves not only the skin and subcutaneous tissues, but also deep tissues. Changes in deep tissues might lead to stretching, angulation, or compression of masticatory nerves, in particular the nerves supplying the masseter and temporal muscles, followed by focal demyelination.^{9,10} Similarly, injury to the motor fibers of the trigeminal nerve from deep tissue changes caused by localized scleroderma might be the reason for association of this condition with HMS.⁴

Electrophysiological studies revealed bursts of high frequency EMG activity and recruitment of synchronized motor unit action potentials, recorded from both masseter muscles and left temporalis muscle. Also, there was no silent period recordable on testing for the masseter inhibitory reflex, on both sides. Till date there is no case report of HMS with evidence of bilateral involvement.

Being a rare disorder, HMS is often confused with other facial movement disorders like hemi facial spasm, oro mandibular dystonia, facial myoclonus, facial tics or other causes of abnormal facial movements. Involuntary clonic tonic movement of muscles innervated by facial muscles characterizes hemi facial spasm and distribution of affected muscles varies according to involved branch.¹¹ As opposed to HMS facial muscles are not hypertrophied in hemi facial spasm. Oro mandibular dystonia is characterized by bilateral repeated sustained involuntary contractions involving lower face.¹² Brief and paroxysmal spasms help to differentiate HMS from unilateral oro mandibular dystonia.

Though pathophysiology is not well known, peripheral trigeminal nerve lesion is considered to play a role.¹⁻⁵ Ectopic discharge secondary to focal demyelination of the trigeminal motor fiber caused by compression, entrapment, or stretching injury to the extracranial portion of the nerve are

all proposed mechanisms.¹⁻³ Vascular compression of the trigeminal motor nucleus or the motor root near the brainstem and hyper excitability of neurons following pontine infarction were the other mechanisms postulated.^{6,8} The exact cause for compression of these distal branches is not known. Mechanical compression due to soft tissue changes as a consequence of atrophy of facial muscles or due to scleroderma seems a distinct possibility. Whether this mechanism could also hold true for asymmetric albeit bilateral involvement of the distal motor branches of the trigeminal nerve is uncertain.⁴

Treatment available for HMS includes oral drugs (carbamazepine, phenytoin and clonazepam), BTX and surgery. BTX has become the treatment of choice now due to excellent outcomes.^{1,3,5-7} The exact role of micro vascular decompression of trigeminal nerve root in patients with HMS is unclear.^{13,14}

In conclusion, HMS is uncommon disorder with unilateral involvement of muscles supplied by trigeminal nerve. In present case clinical and electrophysiological study suggested bilateral though asymmetrical involvement, which is rare. This disabling condition responds very well to botulinum toxin.

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DISCLOSURE

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Conflict of Interest: None

Video 1. [http://neurology-asia.org/content/23/3/neuroasia-2018-23\(3\)-263-v1.mov](http://neurology-asia.org/content/23/3/neuroasia-2018-23(3)-263-v1.mov)
Video showing localized scleroderma involving left forehead, reaching up to vertex. Patient has left facial hemi atrophy and left hemi masticatory spasm involving left masseter and temporalis. Continuous rippling of left half of face, especially over temple and jaw are seen on closure of jaw.

REFERENCES

1. Auger RG, Litchy WJ, Cascino TL, Ahlsgog JE. Hemi masticatory spasm: clinical and electro physiologic observations. *Neurology*1992; 42:2263-6.
2. Cruccu G, Inghilleri M, Berardelli A, et al. Pathophysiology of hemi masticatory spasm. *J Neurol Neurosurg Psychiatry*1994; 57:43-50.

3. Ebersbach G, Kabus C, Schelosky L, Terstegge L, Poewe W. Hemi masticatory spasm in hemi facial atrophy: diagnostic and therapeutic aspects in two patients. *Mov Disord* 1995; 10:504-7.
4. Kim HJ, Jeon BS, Lee KW. Hemi masticatory spasm associated with localized scleroderma and facial hemi atrophy. *Arch Neurol* 2000; 57:576-80.
5. Esteban A, Traba A, Prieto J, Grandas F. Long term follow up of a hemi masticatory spasm. *Acta Neurol Scand* 2002; 105(1): 67-72.
6. Christie C, Rodriguez-Quiroga SA, Arakaki T, Rey RD, Garretto NS. Hemi masticatory spasm: report of a case and review of the literature. *Tremor Other Hyperkinet Mov (NY)* 2014; 4:210.
7. Teive HA, Piovesan EJ, Germiniani FM, *et al.* Hemimasticatory spasm treated with botulinum toxin: case report. *Arq Neuropsiquiatr* 2002; 60:288-9.
8. Gunduz A, Karaali-Savrun F, Uluduz D. Hemi masticatory spasm following pontine infarction. *Mov Disord* 2007; 22(11): 1674-5.
9. Bruyn GW, Bruyn RPM. Hemiatrophies and hemihypertrophies. In: de Jong JMBV, ed: Handbook of clinical neurology, vol. 15. Amsterdam: Elsevier, 1991:475-86.
10. Delaire J, Lumineau JP, Mercier J, Plenier V. Syndrome de Romberg. Hemi-atrophie faciale progressive. *Rev Stomatol ChirMaxillofac* 1983; 84:313-21.
11. Wang A, Jankovic J. Hemi facial spasm: clinical findings and treatment. *Muscle Nerve* 1998; 21(12):1740-7.
12. Sinclair CF, Gurey LE, Blitzer A. Oromandibular dystonia: long-term management with botulinum toxin. *Laryngoscope* 2013; 123(12):3078-83.
13. Sinha R, Chattopadhyay PK, Roychowdhury SK, Bennur S. Hemimasticatory spasm: a case report with a new management strategy. *J Maxillofac Oral Surg* 2011; 10(2): 170-2.
14. Wang YN, Dou NN, Zhou QM, *et al.* Treatment of hemi masticatory spasm with micro vascular decompression. *J Craniofac Surg* 2013; 24(5): 1753-5.