Hemimasticatory Spasm Associated With Localized Scleroderma and Facial Hemiatrophy

Ho Jin Kim, MD; Beom S. Jeon, MD, PhD; Kwang-Woo Lee, MD, PhD

Objectives: To report a case and discuss the mechanism of hemimasticatory spasm.

Design: Case report.

Patient: A 37-year-old woman had a 3-year history of involuntary spasms of the right masseter muscle in association with localized scleroderma and facial hemiatrophy. Electrophysiological studies revealed a normal blink reflex. However, the masseter reflex and silent period were absent on the affected side. Distal latency and compound muscle action potential of the masseter nerve were normal. Needle electromyography demonstrated irregular bursts of motor unit potentials similar to those described in hemifacial spasm. A magnetic resonance imaging scan of the head showed mild hypertrophy of the masseter muscle and atrophy of subcutaneous fatty tissues on the affected side. Local injection of botulinum toxin A into the masseter muscle resolved the patient's symptoms.

Conclusion: On the basis of clinical and electrophysiological findings, focal demyelination of motor branches of the trigeminal nerve owing to deep tissue changes is suggested as the cause of abnormal excitatory electrical activities resulting in involuntary masticatory movement.

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Hemimasticatory spasm (HMS), which is characterized by paroxysmal involuntary contractions of the unilateral jaw-closing muscles, is a very rare disorder of the trigeminal nerve. Initially described as “masticatory spasm of Romberg” by Gowers1 in the last century, to our knowledge only 14 cases of HMS have been reported.2-11 Hemimasticatory spasm usually occurs in association with progressive facial hemiatrophy or localized scleroderma, but may be seen without associated diseases. There has been considerable speculation regarding the underlying mechanism that results in HMS with localized muscle hypertrophy and subcutaneous tissue atrophy.

We report a case of HMS associated with localized scleroderma and facial hemiatrophy with electrophysiological data to delineate the underlying pathophysiological mechanism.

REPORT OF A CASE

A 37-year-old woman first noticed abnormal pigmentation and hardening in the right zygomatic area at the age of 29 years. Subsequent skin biopsy findings were consistent with the diagnosis of localized scleroderma. When she was 34 years old, she began to experience involuntary painful spasms in her right masseter muscle. The spasms were mild at first. However, they increased slowly in duration and frequency over the years, occurring daily and repetitively. Each episode of spasm was usually brief, lasting for only a few seconds, but sometimes lasting for up to a minute.

Examination showed hemiatrophy of the right side of the face. Results of the trigeminal motor and sensory examinations were normal. The involuntary masticatory muscle contractions consisted of brief twitches to prolonged spasms, and were triggered by chewing. A magnetic resonance imaging scan of the head showed mild hypertrophy of the right masseter muscle and reduced subcutaneous fatty tissues in the right side of the face (Figure 1). Blink reflexes were normal bilaterally. The masseter reflexes elicited by tapping the chin using a reflex hammer and recorded by surface electrodes from the masseter and temporalis muscles.
were absent in the right masseter muscle. The silent period after chin tap was also absent in the right masseter muscle (Figure 2). A trigeminal motor nerve conduction study, with needle electrode stimulating below the zygomatic arch and anterior to the temporomandibular joint and surface electrode recording of the masseter muscles, showed normal distal latencies and compound muscle action potential amplitudes bilaterally (Figure 3). Needle electromyography (EMG) of the masseter muscle demonstrated no abnormal spontaneous activity during the resting state. However, during periods of involuntary spasms, it showed brief bursts of motor unit potentials with normal shape discharging at high frequencies up to 200 Hz (Figure 4). The duration of the bursts was variable, lasting from 100 to 600 milliseconds. There was considerable variation in burst intervals, from 50 to 800 milliseconds. These patterns of involuntary EMG activity were triggered by voluntary clenching of the teeth and electrical or mechanical stimulation of the mentalis muscle.

The patient was treated with 20 U of botulinum toxin type A (Botox) injected locally into the right masseter muscle. The spasms disappeared almost completely for 3 months. A subsequent injection was similarly effective in resolution of her symptoms.

**COMMENT**

Our patient showed typical clinical and electrophysiological findings previously described as HMS. Since Kaufman\(^2\) first described the electromyographic characteristics of HMS in 1980, a total of 15 patients (including ours) have been described as having HMS supported by EMG...
findings (Table). A review of these cases suggests that HMS is predominant in women (female-male ratio, 2:1), with a mean age of 31 years (age range, 15-57 years) and a frequent association with facial hemiatrophy (73%) and localized scleroderma (40%).

The involuntary masticatory movement consists of brief twitches or more prolonged spasms. Not all the masticatory muscles are equally involved (Table), and jaw-opening muscles are never affected. Hypertrophy of the involved muscle commonly occurs. Spasms are often exacerbated by forceful voluntary jaw closing and relieved by voluntary jaw opening. It is usually very painful, probably because of excessive contractions of muscle. Sometimes the spasms are so intense that patients may bite their tongue and lips, dislocate their temporomandibular joint, or even break their teeth. Other than the involuntary masticatory movement and hypertrophy of the involved muscle, the findings of neurological examination, including facial sensation and strength, are normal.

Differential diagnoses of HMS include mechanical or inflammatory disorders of the mandible or temporomandibular joint, cephalic tetanus, focal motor epilepsy, unilateral jaw dystonia, tonic spasms of multiple sclerosis, and hemifacial spasm (HFS). Although the clinical features are usually distinct enough to differentiate them from HMS, sometimes it is not easy to differentiate them clinically. However, clinical ambiguities can be easily resolved by electrophysiological studies.

The characteristic EMG findings of HMS are irregular bursts of motor unit potentials lasting from a few seconds to minutes, which correlate clinically with the involuntary twitches or spasms of the masticatory muscles. Individual motor unit potentials are normal and fire at high frequencies up to 200 Hz. These EMG findings of HMS are very similar to those described in HFS.

The high-frequency motor unit discharges in HMS are not present in lesions involving the central nervous system proximal to the motor nuclei and support a peripheral source for the abnormal electrical activity.

Summary of Reported Cases With Hemimasticatory Spasm Supported by Electromyographic (EMG) Findings

<table>
<thead>
<tr>
<th>Source, y</th>
<th>Sex/Age at Onset</th>
<th>Involved Muscles</th>
<th>Facial Hemiatrophy</th>
<th>Electrophysiological Study of Trigeminal Function</th>
<th>EMG Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>F/25</td>
<td>L masseter</td>
<td>Yes</td>
<td>ND</td>
<td>Mild chronic denervation; bursts of 1-4 morphologically different potentials up to 200 Hz</td>
<td></td>
</tr>
<tr>
<td>F/57</td>
<td>R masseter</td>
<td>Yes</td>
<td>ND</td>
<td>Polyphasic potentials of variable amplitude; no clear-cut myogenic or neurogenic signs; cramp recorded from the masseter muscle</td>
<td></td>
</tr>
<tr>
<td>F/57</td>
<td>L temporalis and masseter</td>
<td>No</td>
<td>Normal blink reflexes</td>
<td>Multiple units of normal morphological discharges at high frequencies (20-500 ms)</td>
<td></td>
</tr>
<tr>
<td>F/31</td>
<td>R jaw-closing</td>
<td>Yes</td>
<td>Normal blink reflexes; delayed masseter reflex</td>
<td>20- to 400-ms bursts of motor unit discharge</td>
<td></td>
</tr>
<tr>
<td>F/38</td>
<td>R masseter</td>
<td>Yes</td>
<td>ND</td>
<td>Rapid discharges of high-frequency motor units</td>
<td></td>
</tr>
<tr>
<td>F/34</td>
<td>R masseter</td>
<td>Yes</td>
<td>Normal blink reflexes; silent periods; and MIR; and delay of direct motor response</td>
<td>50- to 100-ms bursts of high-frequency multiple motor unit discharges and recruitment of synchronized motor units leading to 60-Hz tonic activity lasting up to 2 min</td>
<td></td>
</tr>
</tbody>
</table>

* L indicates left; R, right; ND, not described; MIR, masseter inhibitory reflex; and CNS, central nervous system.
Almost a unique finding in HMS.16 However, Ebersbach of silent periods in 1 or 2 muscles on 1 side of the face is voluntary spasm. Even though it may be found in other HMS is the loss of silent periods during episodes of in-
contraction. In contrast to patients with unilateral jaw dystonia, those with involuntary spasms in the orofacial muscles are more prevalent in patients with HMS.17,18

Several hypotheses have been put forward to explain the mechanism of involuntary spasm. These include the following:

1. Loss of inhibition of masseter muscle contraction as a result of ectopic excitation may play a major role, as suggested by the loss of silent periods during periods of involun-
   tary spasms. As to the location of the lesion, some authors have suggested the central nervous system, sympathetic ganglia, or muscle.3,6 However, the electrophysi-
   ological findings argue against these locations. Others have postulated that HMS originates from either the motor root or the motor nucleus of the trigeminal nerve in a similar manner to the proposed mechanism of involuntary activity in HFS.7,8 The EMG findings of HMS, which are very characteristic and quite similar to those of HFS, support this postulation. In HFS, involuntary spasms start in the oribularis oculi and usually spreads to most of the muscles innervated by the facial nerve. In HMS, how-
   ever, involuntary spasms are selectively confined to the masseter, temporalis, and, occasionally, medial ptery-
   goid muscles, while the lateral pterygoid muscle remains spared. Thus, spasms are likely to arise from the distal nerve branches, where fascicles are more widely separated by perineural tissue, not from the motor root or intracranial portion of the mandibular nerve, where the motor fascicles are closely grouped.

Considering the common association of facial hemiatrophy, it has been suggested that deep tissue changes due to facial hemiatrophy cause the trigeminal neuropathy.9 A trigeminal nerve branch to the temporalis and mas-

eter muscles runs in a confined space between the lateral pterygoid muscle and the unyielding surface of the skull, where it might be easily compressed or stretched by deep tissue changes, resulting in focal demyelination of that branch. This anatomical characteristic would explain why the masseter and temporalis muscles are most commonly involved in HMS. It seems unlikely that tri-

geminal neuropathy comes from vascular compression, because, to our knowledge, it has never been docu-

mented at surgical exploration.2,8 Some patients with HMS have connective tissue diseases. However, mononeu-
ropathy associated with connective tissue diseases is an unlikely cause, because the electrophysiological find-

ings of HMS indicate demyelination rather than axonal loss. Furthermore, trigeminal neuropathy associated with connective tissue disease is usually sensory neuropathy but not motor neuropathy.17

On the basis of clinical and electrophysiological find-

ings, HMS and HFS are similar disorders that result from ectopic excitation due to focal demyelination, even though the site and cause of ectopic excitation are different; ie, HFS is caused by vascular compression of the facial nerve near the nerve’s entry into the brain stem, whereas HMS

<table>
<thead>
<tr>
<th>Other Associated Findings</th>
<th>Proposed Site of Lesion and Mechanism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent</td>
<td>Peripheral nerve</td>
</tr>
<tr>
<td>Multiple localized scleroderma</td>
<td>Muscle and CNS</td>
</tr>
<tr>
<td>Absent</td>
<td>Inflammatory disorder or vascular compression of peripheral nerve</td>
</tr>
<tr>
<td>Morphea</td>
<td>Peripheral nerve</td>
</tr>
<tr>
<td>Linear scleroderma</td>
<td>Sympathetic cervical ganglia</td>
</tr>
<tr>
<td>Hypertrophy of L masseter muscle</td>
<td>Peripheral nerve; vascular compression</td>
</tr>
<tr>
<td>Hypertrophy of R masseter and temporalis muscles</td>
<td>Motor root or nucleus</td>
</tr>
<tr>
<td>Absent</td>
<td></td>
</tr>
<tr>
<td>Hypertrophy of R temporalis muscle</td>
<td>Focal demyelination of peripheral muscle nerve by compression</td>
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<tr>
<td>Hypertrophy of L temporalis muscle</td>
<td></td>
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<tr>
<td>Hypertrophy of R masseter and temporalis muscles</td>
<td>Absent</td>
</tr>
<tr>
<td>Linear scleroderma; hypertrophy of L masseter and temporalis muscles</td>
<td>Entrapment of peripheral nerve or hyperexcitability of motor nucleus</td>
</tr>
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<td>Localized scleroderma; hypertrophy of R masseter and temporalis muscles</td>
<td></td>
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<td>Localized scleroderma; hypertrophy of R masseter muscle</td>
<td>Focal demyelination of peripheral nerve by deep tissue damage</td>
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The most diagnostic electrophysiological finding of HMS is the loss of silent periods during episodes of involun-

tary spasm. Even though it may be found in other conditions, such as cephalic tetanus, the complete loss of silent periods in 1 or 2 muscles on 1 side of the face is almost a unique finding in HMS.16 However, Ebersbach et al11 describe a patient with HMS who had no abnor-

ormalities in masseter reflexes or silent period.

The trigeminal motor nerve conduction study per-
formed on our patient showed no abnormalities, includ-

ing distal latencies and compound muscle action poten-

tial amplitudes. Thus, the responsible lesion appeared to be proximal to the stimulation point. One study showed slowing of motor conduction in the infratemporal fossa.9 Both studies showed no reduction in amplitude of the M waves and EMG findings of denervation. These findings indicate demyelination with sparing of the axon.

The mechanisms leading to HMS are still unclear. Impaired inhibition of masseter muscle contraction as a result of ectopic excitation may play a major role, as sug-

gested by the loss of silent periods during periods of in-

voluntary spasm. As to the location of the lesion, some authors have suggested the central nervous system, sympathetic ganglia, or muscle.3,6 However, the electrophysi-

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is caused by compression or stretching of distal mandibular nerve branch owing to deep tissue changes, such as facial hemiatrophy.

Although a few patients have been helped by carbamazepine or phenytoin therapy, treatment with oral drugs has been of no benefit in most patients with HMS. Local injection of botulinum toxin A into affected muscles may be the treatment of choice in HMS.

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REFERENCES