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BRIEF COMMUNICATION

Long-Term Outcome of Hemimasticatory Spasm

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ABSTRACT

Objective This study aims to identify the demographic, clinical, and therapeutic characteristics of four patients with hemimasticatory spasm (HMS) seen in our outpatient department over a period of 20 years.

Methods We performed a retrospective chart review of four patients with HMS who visited outpatient services in the Department of Neurology from 2001 to 2020.

Results The follow-up for all patients ranged from 2 years to 9 years. Three patients had facial or bucco-oral morphea. Two patients maintained long-term improvements in symptoms after being treated with botulinum toxin for 4–7 years, while one patient reported improvement in symptoms with treatment of carbamazepine that subsequently remitted after pregnancy.

Conclusion This report highlights the long-term outcome of HMS in our patients. Our patients reported a significant reduction or complete resolution of symptoms after treatment, and eventually, two patients were asymptomatic while off treatment.

Keywords Botulinum toxin; Spasm; Outcome; Morphea.

Hemimasticatory spasm (HMS) is a rare movement disorder characterized by unilateral, involuntary, and paroxysmal contractions of the muscles of mastication.[1] Spasms are aggravated upon chewing, exposure to low temperature and voluntary closure of the jaw.[1,2] The medial pterygoid, masseter, and temporalis muscles are affected by this condition.[3]

HMS has been largely implicated in peripheral motor disorder. The selective involvement of the masseter and temporalis muscles indicates affliction of the distal mandibular nerve rather than involvement at the motor root.[1] The involvement of trigeminal motor fibers in the form of focal demyelination and ectopic impulse generation has been one of the proposed mechanisms.[1,3,4] Because of the rarity of the disease, the long-term outcome of HMS has seldom been studied. In a previous report, one patient with HMS had a chronic illness, requiring regular botulinum toxin (BT) administration for 11 years.[5] Given the rarity of illness, HMS is frequently misdiagnosed as oromandibular dystonia and hemifacial spasm.[3,5] We describe the demographic, clinical, and therapeutic characteristics of four patients with HMS seen in our outpatient department over a 20 year period.

MATERIALS & METHODS

We performed a retrospective chart review of four patients with HMS who visited the outpatient department and Movement Disorders Clinic of the Department of Neurology at the National Institute of Mental Health and Neurosciences (NIMHANS), Bangalore, India from 2001 to 2020. The study was approved by the Institute Ethics Committee of NIMHANS (No. NIMH/DO/DEAN [Basic Science]/2020-21). All subjects gave written informed consent to participate in the study. All procedures were conducted in accordance with the ethical standards of the Institutional Ethics Committee and with the 1975 Helsinki declaration.

RESULTS

Patient 1

A 24-year-old man presented with painless involuntary abnormal movements of the left cheek and lateral aspect of the head

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for 3 years. He was diagnosed with morphea (localized scleroderma) on the left cheek at the age of 13 years and underwent 8 sessions of laser treatment at 2–3 month intervals.

Upon examination, there was localized hyperpigmentation over the left cheek and face associated with depressed scars (Figure 1A). Involuntary movements of the left masseter and temporalis muscles were observed that disappeared during jaw movement.

The patient was treated with 0.75 mg of clonazepam per day. In addition, 100 units (U) of BT were administered to the left masseter with good results. The patient was on regular BT injection for 7 years. Subsequently, BT therapy was deferred for the next 2 years as a result of mild spasms with a frequency of 1–2/month.

Patient 2

A 31-year-old woman presented with a seven-year history of sudden episodes of teeth clenching, as well as contraction and stiffening of the right side of the face that was associated with severe pain and occasional tongue bites. The patient recovered spontaneously from the attacks. Each episode initially lasted for 10–20 seconds, and gradually increased to one minute. Symptoms occurred at an interval of 15 days to one month, at a rate of 1–2 times per day. The frequency of episodes progressively increased during pregnancy, particularly after the seventh month of pregnancy. Symptoms were aggravated by stress, cold climate, and fasting. The spasms ceased in the eighth month of pregnancy after being treated with 400 mg of carbamazepine per day. Brain MRI showed the presence of a vascular loop adjacent to the facial nerve, and the area around the trigeminal nerve was normal.

Upon examination, right hemifacial atrophy was noted. Evaluation by a dermatologist revealed bucco-oral morphea. The patient was not treated with BT, as she was five months postpartum and was lactating. After taking carbamazepine for four months, the patient stopped the medication, and she continued to remain asymptomatic for 2 years.

Patient 3

A 42-year-old man presented with painless involuntary movements of the right half of the face for one year that worsened over the past 4–5 months, which was associated with occasional cheek and tongue bites. The movements were induced by opposition of both jaws and worsened while talking. The movements were suppressed when he opened his mouth.

He also complained of swelling of the left side of his neck for 3 years. Six months after the onset of muscle spasms, he was diagnosed with Hodgkin's lymphoma and was initiated on chemotherapy. He was started on clonazepam, carbamazepine, and baclofen for the treatment of spasms, with which he reported approximately 50% improvement. He was also treated with BT but reported no benefit.

Upon examination, rippling movements of the right masseter muscle were observed upon clenching the teeth (Supplementary Video 1 in the online-only Data Supplement). The bulk and power of the masseter and opening and closing movements of the jaw were normal. One year after diagnosis, examination revealed hypertrophy of the right temporalis and masseter muscles. MRI showed the superior cerebellar artery compressing the right trigeminal nerve (Figure 1B). The patient was advised to undergo a repeat trial of BT, but he declined. At a 2 year follow-

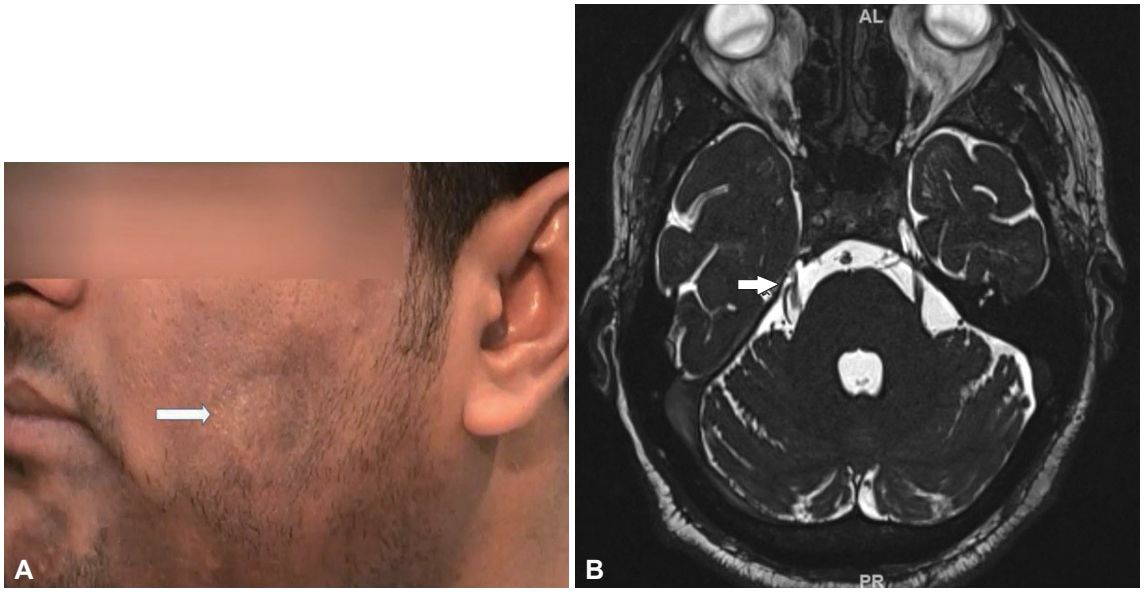

Figure 1. Clinical and radiological features of hemimasticatory spasm. A: Morphea (arrow) on the left cheek of Patient 1. B: MRI (cis 3D sequence) shows a vascular loop (superior cerebellar artery, arrow) around the right trigeminal nerve of Patient 3.

up, he continued to have symptoms that improved by approximately 30% with the medications.

Patient 4

A 37-year-old woman presented with a history of episodic painful spasms of jaw muscles (mainly left) with a duration of 1 year and 2 months. The onset of episodes were sudden and occurred while talking and chewing. She began treatment with carbamazepine, clonazepam and gabapentin, and the doses were gradually increased (carbamazepine 1,200 mg/day, clonazepam 4 mg/day, gabapentin 1,200 mg/day) with which the spasms were reduced. Two months after the onset of spasms, she developed thinning of the skin over the scalp, neck, back and abdomen, which is suggestive of morphea. She received prednisolone 2 mg/kg 2 times/week for two months and chloroquine with no benefit. However, there was improvement in morphea with placental extract.

Upon examination, she had scars over the face, left side of the neck and back along with hypertrophy of the left masseter muscle. While speaking, she had sudden jaw muscle spasms that caused jaw closure. Imaging results showed a vascular loop (superior cerebellar artery) around the left trigeminal nerve. The patient was treated with BT injections of 30−35 U in the left masseter and 15 U in the right masseter, with which the symptoms resolved.

At follow-up after 5 years, the patient's symptoms had improved significantly while on BT injection, clonazepam, and gabapentin. She had taken BT injections for 4 years and then discontinued without any recurrence of symptoms (Table 1).

DISCUSSION

In this case series, we described four patients (2 women) with HMS, which is an extremely rare clinical condition. To date, only 65 cases have been reported in the literature (Supplementary Table 1 in the online-only Data Supplement).[2,6,7]

Patient 1 had significant improvement of spasms after being treated with BT injection. The retrograde transport of BT leading to modulation of central inhibitory circuits could explain recovery following prolonged BT administration.[8] In a previously reported case, although the improvement of symptoms with BT was dramatic, the patient needed regular BT injections to maintain remission of symptoms.[9] The improvement of symptoms in this patient could also be due to the self-limiting nature of the illness rather than the effect of BT.

Patient 2 reported exacerbation of symptoms during the seventh month of pregnancy that eventually improved during the postpartum period. A similar case has been previously reported.[10] The cause for complete resolution of symptoms in patient 2 could be due to the probable self-limiting nature of the illness, and morphea could just be an association rather than an etiology. The modulatory effects of hormonal surge during the postpartum period could be another plausible mechanism; however,

Table 1. Clinical features and follow-up of patients with HMS

Patient number	AAO/sex	Clinical features	Coexisting illness	MRI brain	Treatment	Last follow up	Follow up details
1	21/male	Involuntary movements of left masseter, temporalis	Morphea diagnosed at 13 years	Not done	BT	9 years after first presentation	Patient has been regularly on BT until 2 years back Markedly reduced frequency of spasms
2	24/female	Sudden clenching of right jaw and teeth	Exacerbation during 7th month of pregnancy Diagnosed with bucco-oral morphea	AICA vascular loop but not compressing facial nerve	Carbamazepine	2 years after first presentation	Spontaneous remission of symptoms for 1 year
3	39/male	Involuntary movements of right half of face Examination revealed rippling movements of right masseter	Diagnosed with Hodgkin's lymphoma 2.5 years after onset of HMS	Superior cerebellar artery compressing right trigeminal nerve	Carbamazepine, clonazepam, baclofen	2 years after first presentation	Improvement in symptoms by 30%
4	36/female	Episodic spasms of jaw muscles Examination revealed left masseter involvement	Morphea diagnosed at the age of 36 years	Vascular loop around right trigeminal nerve	BT, clonazepam, gabapentin	5 years after first presentation	Spontaneous remission of symptoms after 4 years of BT

AAO, age at onset; BT, botulinum toxin; AICA, anterior inferior cerebellar artery; HMS, hemimasticatory spasm.

there is insufficient evidence to support this hypothesis.

Patient 3 was diagnosed with Hodgkin's lymphoma a few months after the onset of jaw spasms.

HMS has been commonly associated with morphea[1,2] and Parry-Romberg syndrome.[6,11] The association with morphea suggests that skin sclerosis might be a predisposing factor in narrowing the space between the medial pterygoid and the mandibular bone, leading to compression of the temporomasseter nerve. Atrophy and depigmentation on the chin have been observed in patients with HMS who also had involuntary lateral jaw movements due to involvement of the lateral pterygoid.[6,12]

The presence of a vascular loop around the trigeminal nerve in patients with HMS and subsequent relief of symptoms following microvascular decompression has been reported.[13-16] Removal of the masseter nerve root attached to the temporomandibular joint has shown favorable outcomes.[7] Debulking of the masseter muscles has also yielded good improvement.[17]

The exact pathophysiology of HMS is still not clear. HMS is usually precipitated by talking, laughing, etc. The lack of sensory processing differentiates it from hemifacial spasm.

In patients 1 and 2, the HMS could have originated from the focal motor branch of the postganglionic trigeminal nerve (presence of morphoea), and in the other two patients, there could be preganglionic involvement of the trigeminal nerve (superior cerebellar artery abutting the trigeminal nerve). Electrophysiological studies have shown the involvement of trigeminal motor fibers in the form of focal demyelination and ectopic impulse generation. According to a report, HMS should be considered within the spectrum of facial spasms.[18]

Suppression of HMS by slightly opening the mouth has been reported earlier.[8] Electromyographic recording of muscles during HMS is similar to that of a cramp. Voluntary jaw closure is known to trigger HMS, and by opening the mouth, the masseter is lengthened, which leads to inhibition of HMS, similar to what is seen with cramp abolition.[19]

In conclusion, this case series adds to the existing literature regarding HMS. Follow-up in these patients has highlighted the benefit of long-term administration of BT in complete alleviation of symptoms, indicating a favorable prognosis in these patients.

Supplementary Video Legends

Video 1. Right hemimasticatory spasm on clenching of the teeth.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.21067.

Conflicts of Interest

The authors have no financial conflicts of interest.

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Author Contributions

Conceptualization: Pramod Kumar Pal. Data curation: Somdattaa Ray. Formal analysis: Somdattaa Ray. Investigation: Somdattaa Ray, Nitish Kamble. Methodology: Ravi Yadav, Nitish Kamble, Pramod Kumar Pal. Project administration: Somdattaa Ray, Nitish Kamble. Resources: Nitish Kamble, Pramod Kumar Pal. Supervision: Nitish Kamble, Pramod Kumar Pal. Validation: Nitish Kamble. Visualization: Somdattaa Ray, Nitish Kamble. Writing—original draft: Somdattaa Ray. Writing—review & editing: Nitish Kamble, Ravi Yadav, Pramod Kumar Pal.

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Supplementary Table 1. Summary of follow-up and outcome details from previous studies in HMS

Sl. No.	Study	Clinical features	Treatment given	Last follow up	Outcome at last follow up
1	Auger et al.[3]	Patient 1: 25-year-old lady with right masseter and temporal muscle painful spasms	Patient 1: sectioning of trigeminal motor rootlets followed by BT	Patient 1: not available	Patient 1: significant improvement
		Patient 2: 31-year-old female with right medial pterygoid spasm	Patient 2: carbamazepine	Patient 2: not available	Patient 2: significant improvement
		Patient 3: 25-year-old lady presented with involuntary spasms of right temporalis and masseter	Patient 3: no treatment was given	Patient 3: 15 years	Patient 3: no change in symptoms
2	Cruccu et al.[4]	Patient 1: 20-year-old male presented with involuntary twitches of left temporalis muscle	Patient 1: carbamazepine 1,200 mg/day	Patient 1: 6 months	Patient 1: moderate benefit
		Patient 2: 55-year-old woman presented with spasms over right masseter and temporalis muscles	Patient 2: carbamazepine 600 mg/day, BT 30–50 U	Patient 2: not available	Patient 2: significant improvement
3	Esteban et al.[5]	61-year-old lady presented with involuntary left masseter spasm	Not mentioned	11 years	Clinical course remained unchanged
4	Zhang et al.[7]	Two patients presented with HMS	Avulsion of masseter nerve	3 years	Complete resolution of symptoms
5	Sun et al.[13]	22-year-old male with left masseter spasm	Vascular decompression of trigeminal nerve	12 months	Complete resolution of symptoms
6	Wang et al.[15]	6 patients with HMS	Microvascular decompression of motor branch of trigeminal nerve	4–58 months	Complete resolution of symptoms
7	Dou et al.[16]	56-year-old female with spasm of left masseter	Microvascular decompression of trigeminal motor rootlet	7 months	Complete resolution of symptoms
8	Sinha et al.[17]	38-year-old male with involuntary spasm of right masseter and temporalis muscle	Debulking and stripping of masseter muscles	1 year	Complete resolution of symptoms
9	Present study	4 patients (2 men and 2 women) with HMS	2 received BT and 2 were on medications	2–9 years	Complete resolution in 2 and partial resolution in 2 patients

HMS, hemimasticatory spasm; BT, botulinum toxin.