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Effectiveness of Botulinum Toxin A for Treatment of Hemimasticatory Spasm

[1]Dr. Abhishek Singh, M.D.S., Senior Resident, Department of Oral and Maxillofacial Surgery, AIIMS, Patna

[2]Dr Sailesh Kumar Mukul, M.D.S. Associate Professor, Oral and Maxillofacial Surgery, AIIMS, Patna

[3]Dr Sumit Singh, M.D.S., Senior Resident, Prosthodontics Crown and Bridge & Implantology, AIIMS, Patna

[4]Dr Dhritiman Pathak, M.D.S. Oral and Maxillofacial Surgery, Consultant Maxillofacial Surgeon, Sarvodaya Healthcare, Guwahati, Assam

[5]Dr. Aiswarya Vaidyanathan, M.D.S., Junior Resident, Oral and Maxillofacial Surgery, AIIMS, Patna

Corresponding author: Dr. Abhishek Singh, M.D.S., Senior Resident, Department of Oral and Maxillofacial Surgery, AIIMS, Patna

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Abstract

Hemi masticatory spasm (HMS) represents a rare facial movement disorder that is characterized by paroxysmal involuntary contractions of one or more of the jaw-closing muscles (masseter, temporalis, and medial pterygoid) on one side of the face due to a dysfunction of the motor branch of trigeminal nerve with unknown etiology. Various treatment options for HMS are the use of oral drugs, surgical treatment and injection of botulinum toxin. In this study, a case of HMS was discussed with masseter muscle hypertrophy which was treated with botulinum toxin A, which was found to be most effective in reducing the size of hypertrophied muscle and should be considered in the spectrum of facial spasm.

Keywords- Hemi masticatory spas (HMS), Hemifacial Spasm (HFS), Botulinum toxin A, Electromyography (EMG)

Introduction

Hemi masticatory spasm (HMS) represents a rare facial movement disorder that is characterized by paroxysmal involuntary contractions of one or more of the jaw-closing muscles (masseter, temporalis, and medial pterygoid) on one side of the face due to a dysfunction of the motor branch of trigeminal nerve with unknown etiology. It is frequently misdiagnosed as hemifacial spasm (HFS), which is a disorder due to dysfunction of the facial nerve (1-4)

It was first described by Gowers in 1897, who named it as 'masticatory spasm of Romberg'(5), due to its rarity, HMS may be confused with other facial movement

disorders, such as hemifacial spasm (HFS), temporomandibular joint disorder (TMD), oromandibular dystonia (OMD), facial myoclonus, facial tics, or other causes of abnormal facial movements(6).

There are many treatment options for HMS, ranging from medical to surgical approaches. A milestone in the treatment of HMS is botulinum toxin, which has become the treatment of choice due to its excellent results. Here we report a case of HMS which was successfully treated with botulinum toxin A.

Case Presentation

A 49 year old male patient reported to the department with a history of recurrent spontaneous spasm, involuntary contraction of jaw with sporadic injuries to the tongue and strange sensation in the preauricular region. A detailed history of the patient was obtained which revealed that he had bitten his tongue many times during the episodes of the spasm which had gradually increased with time reaching upto 25 episodes in a day. The episodes consist of sudden twitches in the preauricular region with pain which hamper the movement of jaw for as long as 20 seconds.

On clinical examination mild hypertrophy of masseter muscle was noted. No relevant medical and family history was obtained. The patient was advised for a Computerized Tomography (CT) scan and surface Electromyography (EMG). The CT scan was done to rule out any brain pathology, nerve conduction finding were in normal limits. EMG revealed spontaneous irregular bursts of motor unit potentials (MUPs) in left temporalis and masseter region, suggesting hyperexcitability of these muscles. The patient was administered injections of 50 units of botulinum Type A in the involved muscles. The injections were administered to the patients every 4 months which gave an excellent response, the patient revealed reduction in the spasmodic episodes as well as reduction in pain on the affected side.(Fig-1,2,3)

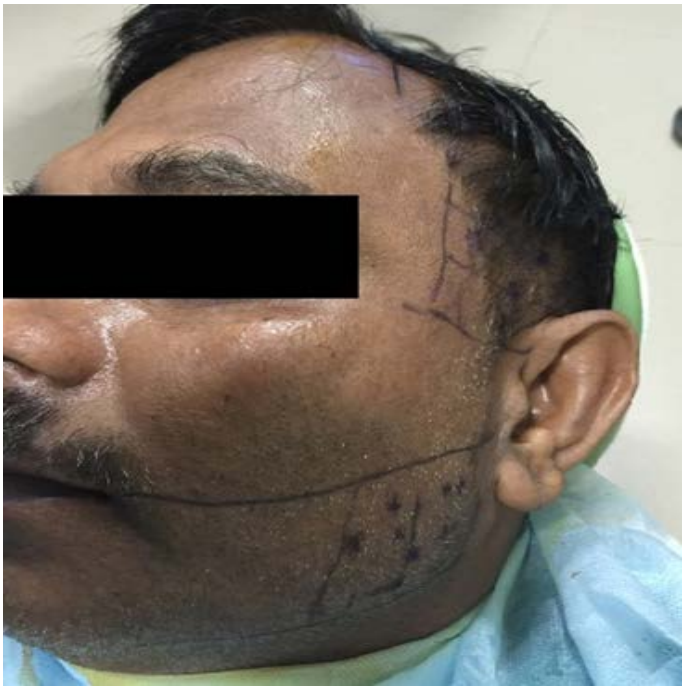

Fig 1: lateral facial diagram of marking and position of injections in temporalis and masseter muscles.

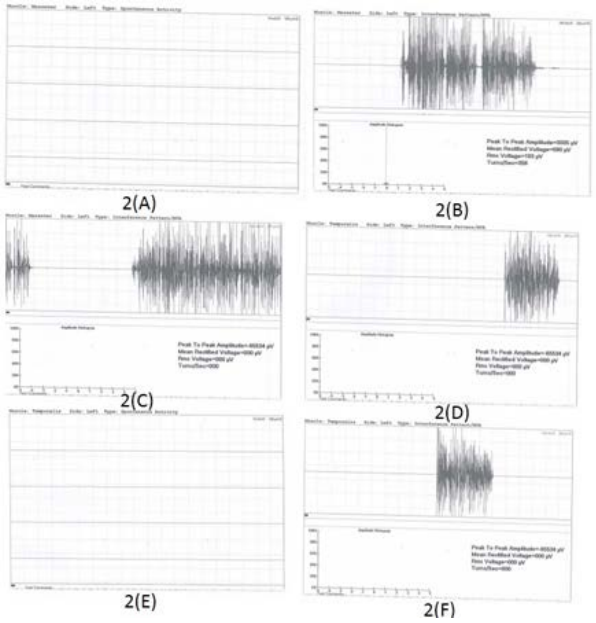

Fig 2 : Surface EMG of left temporalis and masseter muscles showing spontaneous irregular burst of motor unit potential (MUPs)

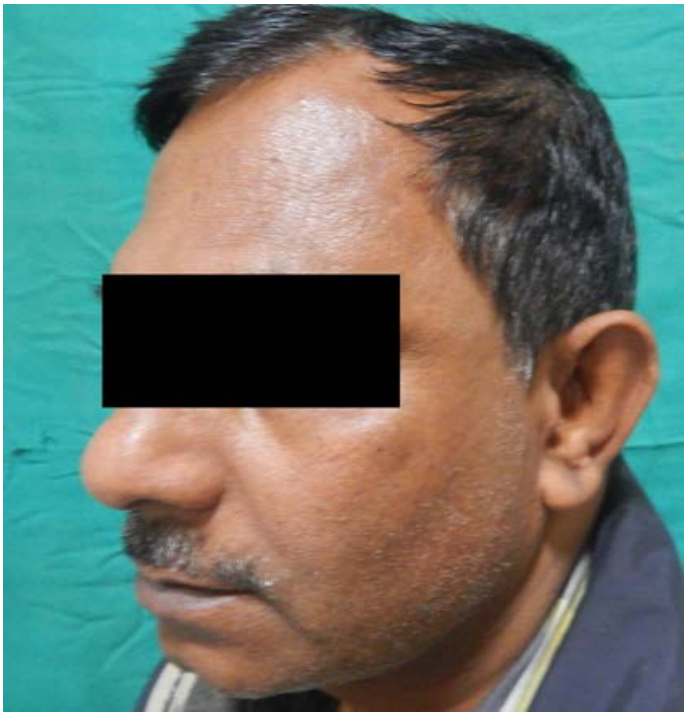

Fig 3: Image after 4 months of Botulinum toxin injections.

Discussion

HMS is a rare movement disorder characterized by paroxysmal contractions of unilateral jaw-closing muscles, such as the masseter, temporalis, and medial pterygoid muscles (3). However, the involvement of jaw opening muscles has never been described (4,7).To our knowledge, only 38 cases of HMS have been reported in the literature. HMS predominantly affects female (female : male = 2.8:1.0), in the third to fourth decades of life (mean age-39 year) and is associated with hemiatrophy in two-thirds and scleroderma in one-third of the cases (4,7,8).

The involuntary muscle contractions usually last from a few seconds to several minutes. The episodes of spasm may occur many times a day, it is often precipitated by chewing, speaking or other voluntary jaw-closing, and may be interrupted by voluntary jaw opening. Occasionally, sudden and severe spasms may result in biting of tongue or oral mucosa, breaking of teeth, or even temporomandibular joint dislocation (1,3,9).

Although the pathophysiology of HMS is not well known, but peripheral lesion of the trigeminal motor nerve is considered to play a role in it (3,4,7-10). It has been proposed that HMS is produced by ectopic discharge, secondary to focal demyelination of the trigeminal motor nerve fiber caused by compression, entrapment, or stretching injury to the extracranial portion of the nerve (3,4,7,8,11,12). Some authors also have postulated that HMS might be originated from vascular compression of the trigeminal motor nucleus or motor root near the brainstem (3,4,7,12).

EMG and MRI are helpful in localization of the pathological lesion so also helping in establishing the diagnosis. Characteristic EMG results for HMS shows rapid burst of motor unit potentials with high frequencies (up to 200 Hz) that correlate with the involuntary spasms of the involving masticatory muscles(mostly affecting the masseter and then the temporalis muscle) (3,4,7), which is seen in our case.

The most diagnostic feature of HMS is the loss of silent periods during the muscle spasm. The silent period is defined as a reflexive pause in muscle activity during functional tooth contact (as in chewing or swallowing), or following an experimentally produced tap on the chin of a subject whose teeth are clenched together (jaw-jerk reflex) (13).

The electrophysiologic findings are characteristic in HMS and help in the differential diagnosis with other conditions like unilateral dystonia of the jaw with jaw closure, temporomandibular joint syndrome, paroxysmal events in multiple sclerosis and tetany (6).

MRI is helpful to determine the secondary causes of HMS from brain lesions which including pontine infarction (14), biopercular syndrome (15), and cerebellopontine angle hematoma22 (16) or lesions along the trigerminal nerve.

Muscle hypertrophy and Subcutaneous tissue atrophy can be easily identified through coronal view of MRI. (7) MRA (Magnetic resonance Angiography) is helpful in clarifying the vascular pathology associated with the trigeminal nerve.

Various treatment options for HMS are the use of oral drugs, surgical treatment and injection of botulinum toxin (3,8). Attempts at treatment with phenytoin, carbamazepine, clonazepam, diazepam, dantrolene, baclofen, valproate, haloperidol, amitriptyline, trihexyphenidyl, and cyclobenzaprine have been tried but not beneficial in most of the patients. However, one of two patients described by Thompson et al (6) shows good clinical improvement improvement while taking carbamazepine and phenytoin.

Auger et al. published a clinical and electrophysiologic observations on 3 cases of hemimasticatory spasm, and one of them responded favorably to botulinum toxin injections (3). Ebersbach et al. reported two cases of hemimasticatory spasm in hemifacial atrophy; injections of botulinum toxin type A into the masticatory muscles proved to be a successful treatment in both patients (8). Kim et al. reported a case of hemimasticatory spasm associated with localized scleroderma and facial hemiatrophy; the authors described excellent results after the use of botulinum toxin type A injections (7).

Botulinum toxin type A into involved muscles may be considered the treatment of choice for the movement disorder and pain in HMS, with doses of 40-100 units in the masseter and temporalis muscles. In addition, Botulinum toxin type A can be helpful for decreasing the size of the hypertrophied muscle.

Conclusion

In this study, a case of HMS was discussed with masseter muscle hypertrophy which was treated with botulinum type A, which was found to be most effective in reducing the size of hypertrophied muscle and should be considered in the spectrum of facial spasm.

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