Case Report

Oro-mandibular dystonia

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ABSTRACT

Dystonia is an involuntary, repetitive, sustained (tonic), or spasmodic (rapid or clonic) muscle contraction. The spectrum of dystonias can involve various regions of the body. Oromandibular dystonia (OMD) can involve the masticatory, lower facial and the tongue muscles which may results in trismus, bruxism, involuntary jaw opening or closure and involuntary tongue movement. Here, we report a case of OMD in a 68 year old man.

Key words: Botulinum toxoid, dystonia, masticatory, musculature, oromandibular

Introduction

Dystonia is an involuntary, repetitive, sustained (tonic), or spasmodic (rapid or clonic) muscle contraction. The spectrum of dystonias can involve various regions of the body. Of interest to the oral and maxillofacial surgeon are the cranial-cervical dystonias and, in particular, oromandibular dystonia (OMD).

Cranial-cervical dystonia is an involuntary, sustained contraction of the periorbital, facial, oromandibular, pharyngeal, laryngeal, or cervical muscles.[1] OMD can involve the masticatory, lower facial and the tongue muscles which may results in trismus, bruxism, involuntary jaw opening or closure and involuntary tongue movement. These involuntary movements may produce inappropriate deviation of the mandible, subluxation, intraoral soft tissue trauma and bone resorption. Dysphagia, dysphonia and difficulty with mastication also often occur with OMD.

OMD is frequently associated with other cranial-cervical dystonias, especially blepharospasm. This combination is referred to as Brueghel's syndrome. Meige syndrome is orofacial dystonia and closely related?

The etiology of OMD is varied and includes genetic predisposition, injury to the central nervous system, peripheral trauma, medications, metabolic or toxic states and neurodegenerative disease. In majority of the patients, no specific cause can be identified.

Some studies suggest that OMD affects women more than men with a mean age of onset of the symptoms between 31 and 58 years. Although there is some evidence that genetic predisposition may be a factor in some OMD patients, in most cases the etiology of OMD is elusive. It is not uncommon for patients to report a precise onset of the first OMD episode.

In studies by Tan and Jankovic, idiopathic etiology accounts for majority (63%) of the cases reported. Other possible etiologies include drug-induced OMD (22.8%), peripheral-induced OMD (9.3%), post-anoxia OMD (2.5%), neurodegenerative disorder associated OMD (1.8%) and head injury-associated OMD (0.8%).

Case Report

A 68-year-old man presented to the oral and maxillofacial surgery out-patient clinic. His chief complaint was inability to maintain the lower jaw in one position and severe muscle spasms that frequently caused severe clenching of teeth on the right side with obvious gnashing sounds due to bruxism. A visit to the local dentist for rehabilitation of worn out teeth resulted in this referral.

He had been suffering from this problem with its intensity increasing gradually for 3 years. Previous treatment had included occlusal equilibration, multiple occlusal appliances and intramuscular injections of local anesthesia.

Access this article online

Quick Response Code:

Website: www.njms.in

DOI: 10.4103/0975-5950.79218

One year back, he was examined by a neurologist for the same problem, who advised investigations including radiographs, magnetic resonance imaging of the brain and right Temporomandibular joint. The patient was kept on a regime of Tetrabenazine 25 mg and Haloperidol 0.75 mg twice daily, along with a nocturnal bite guard. The magnetic resonance image revealed slight periventricular ischemia with hypertrophy of the masseter and coronoid process of right side [Figure 1].

Current examination revealed severe clenching of molars on the right side with slight ipsilateral deviation of the mandible. The left masseter and temporalis musclesdid not show any abnormal excessive contractions as on the right. A working diagnosis of OMD was made since all other modes of therapy failed to provide relief to the patient [Figure 2].

A two pronged treatment plan was arrived at, comprising local injections of Botulinum toxin A[2-4] into the masseter and temporalis muscles, followed by coroniodectomy and medial debulking of the masseter on the right side.

Electromyography guided Botulinum toxin A injections[5] were given as follows:

1. right masseter: 35 units,
2. left masseter: 20 units,
3. right temporalis: 20 units and
4. left temporalis: 15 units.

EMG guidance leads to deposition of the solution in direct vicinity of the hyperactive muscle fibers,[6-8] unlike blind probing and injecting where much of the solution may end up in the vicinity of not so hyperactive fibers [Figures 3 and 4].

A week later, the patient showed a definitive reduction in the frequency of dystonic movements but the clenching force on the right side remained the same.

As planned, the patient was administered general anesthesia and the hypertrophic coronoid was removed on the right side and concomitant debulking of the right masseter was carried out via standard intraoral approach. Two weeks later, the patient was completely

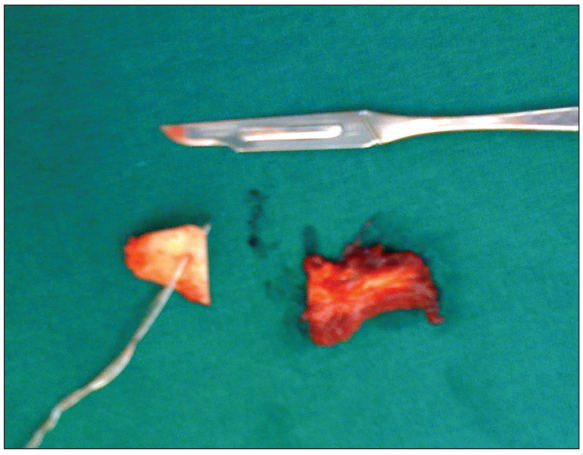

Figure 1: Coronoid and deep fibers of masseter

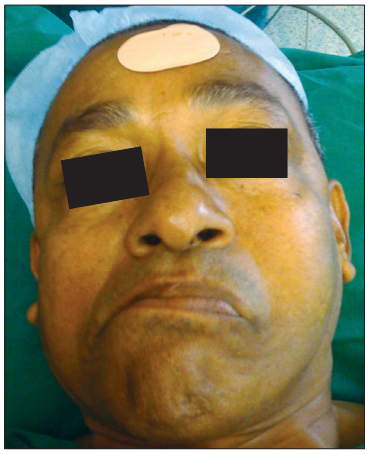

Figure 2: Preoperative frontal view

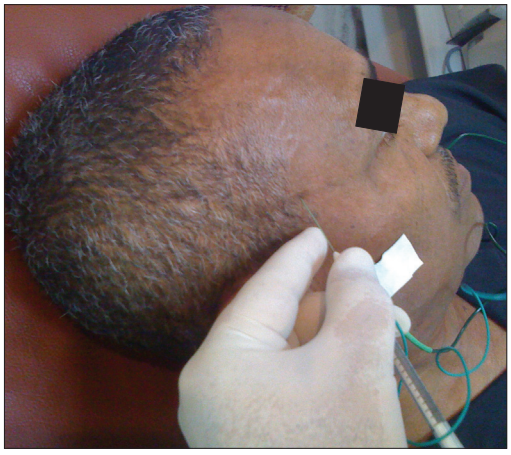

Figure 3: EMG guided botox injection

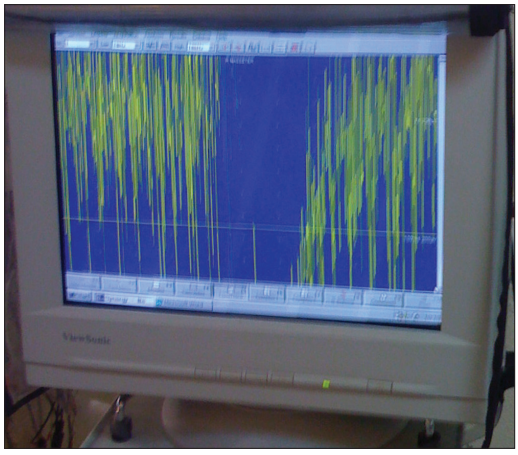

Figure 4: Hyperactive muscle fibers

relieved from the dystonic movements and showed a marked reduction in diurnal clenching activity.

Discussion

The patient in study reported for a checkup after 18 months and upon examination it was found that there was alleviation of symptoms except for minor bruxism on the right albiet less in frequency than before.

For this he was administered a dose of the same botulinum toxoid 15 units each in the right temporalis and masseter respectively.

So we saw a near complete alleviation of symptoms in the above mentioned case with the protocol followed.

Due to the varied clinical presentation of OMD, the differential diagnosis may be very challenging and requires a thorough history and clinical examination including a psychological evaluation. Patients with movement disorders such as OMD may present with accompanying psychiatric conditions such as depression, anxiety, obsessive-compulsive problems, schizoid personality, space phobia and other psychological abnormalities. The psychological profile may further confuse the clinician and confound the diagnosis. Currently, there is no evidence to suggest psychological causes of OMD. However, they can certainly coexist.

A thorough history and examination including a psychological evaluation will safeguard against incorrectly diagnosing extraordinary presentations of OMD as psychological. Once the patient's history and psychological evaluation are completed, the offending muscles need to be localized. It was believed that for this patient, the right masseter and temporalis muscles muscles were responsible for the mouth opening dystonia.

Earlier reviews reported that myotomies offer little benefit and in recent years they have been abandoned. The primary purpose was to abolish abnormal movements in all the muscles involved in producing the movement while preserving innervation of those that are not involved.

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Source of Support: Nil. **Conflict of Interest:** None declared.