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Teemul, TA, Patel, R, Kanatas, A et al. (1 more author) (2016) Management of oromandibular dystonia with botulinum A toxin: a series of cases. British Journal of Oral and Maxillofacial Surgery, 54 (10). pp. 1080-1084. ISSN 0266-4356

https://doi.org/10.1016/j.bjoms.2016.06.028

Management of oromandibular dystonia with botulinum A toxin: a series of cases.

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Introduction

Oromandibular dystonia (OMD) is a movement disorder characterized by involuntary, repetitive, and patterned muscle contractions of varying severity, affecting the jaws, tongue, face and pharynx.[1] It usually manifests as muscle contraction with repetitive movement, abnormal postures and a wide variation in clinical presentation- depending on the musculature affected.[2]
Dystonia can be classified into regions of distribution: Focal, segmental, multifocal and generalized. It can also be primary or seconday based on aetiology (see table 1).

Primary type
Idiopathic
Inherited
Familial with genetic predisposition
Secondary type
Peripheral trauma
Surgical complication
Diseases of the brain
Neurodegenerative disorders
Cerebral infarction
Drug induced

Table 1.

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The most common secondary type is tardive dystonia, which develops as a side effect of long term treatment with antipsychotic drugs. Oro-facial-lingual stereotypes are more frequent in the tardive than the idiopathic group, which is often associated with cervical dystonia.[3]
The clinical characteristic of the dystonia may be classified as jaw opening, jaw closing, jaw deflecting, jaw retruding or a mixed/combination type. The differential diagnoses for OMD include temporomandibular joint disorders, hemifacial spasm and psychological disorders. When OMD occurs with blepharospasm it is known as Meige's syndrome.
Patients have reported triggers such as stress, talking, chewing and praying. There are some learned behaviours that help reduce the dystonia such as sleeping, relaxing, talking, singing, lip biting, tongue posturing, swallowing and chewing.[4]
Oral medication is often the first line of treatment as systemic pharmacologic therapy benefits about 30% of patients.[2] Although most oral medications have a low success rate, treatment includes cholinergics, benzodiazepines, anti-parkinsonism drugs, anti-convulsants, baclofen, carbamazepine and lithium. Anticholinergic medications have been found to be the most effective oral medication for the treatment of dystonia. Muscle afferent block (MAB) has been found to be highly effective for OMD but has no effect in patients with dyskinetic symptoms.[5] MAB involves intramuscular injection of anaesthetic and alcohol and has been used for the treatment of OMD. However, since it is not as effective with dyskinetic symptoms, this suggests that OMD and oral/orofacial dyskinesia may have a different physiopathology.
The next line of treatment is Botulinum toxin injections which are the mainstay of treatment for most focal dystonias. The response rate has been reported as high as 90-95% with Botulinum toxin type A.[6] However, some patients can become secondarily nonresponsive as Botulinum neurotoxins may be immunogenic leading to immuno-resistance.
Jaw closing dystonia responds better than jaw opening or mixed dystonia. The treatment of the latter types of OMD is more likely associated with dysphagia and dysarthria.

Method

A literature review was undertaken. The patients treated for OMD were identified in a specific period from 2013-2015. These cases were analysed with a data collection tool. The outcome was measured using the Glasgow Benefit Inventory (GBI) score. This is a questionnaire used to measure the change in health status produced by the intervention. There are 18 items and it is intended for use on adults- representing a measure of patient-reported outcome. It is a subjective patient oriented post-interventional questionnaire which is sensitive to any change in health status brought about by a specific event- in this case the administration of Botulinum toxin to treat OMD. GBI scores can range from -100 (maximal adverse effect), through 0 (no effect), to 100 (maximal positive effect).

Results

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The results of the literature review are summarized in table 2. One study using the GBI questionnaire showed a mean score of +38.04.[7] Overall, there was a positive beneficial result with Botulinum toxin treatment.
In our experience, there were 6 patients treated for OMD with Botulinum toxin type A. The results are summarized in table 3. The mean dose of toxin delivered to the various muscles: masseter, temporalis, lateral pterygoid, anterior digastric and genioglossus were 20, 15, 24, 15 and 20 units respectively to each side.
The overall GBI score was positive for all except one patient. Further analysis of the GBI score reflected that patients improved in the sub-categories of general well being, social and physical outcomes (see table 3). The mean GBI score was +40. The mean improvement in the categories of general health, social health and physical health were 82.17%, 37.17% and 31.17% respectively.

Author, year	Number of patients	Diagnosis	Treatment given	Outcomes	Complications
Moscovich et al, 2015	8	Open jaw/jaw deviation dystonia	Onabotulinum toxin using internal approach	-6 patients very much improved -2 patients much improved (Global Impression scale)	One adverse event-nasal speech following injection
Jankovic et al, 2015	1046	Cervical dystonia	189.8 ±87.1 Units Botulinum toxin, average treatment interval 14.6 and 15.1 weeks	-Score decreased from 39.2 to 27.1 (Toronto Western Spasmodic Torticollis Rating Scale) -Score improved from 91.2% to 95% (Clinical Global Assessment of change) -Score improved from 83% to 91.7% (Clinical Global Assessment of change) -Reduction in cervical dystonia impact profile	-26.2% reported adverse events -7.0% muscle weakness -6.4% dysphagia
Pedemonte et al, 2015	30	Post traumatic OMD	Onabotulinum Toxin A	Symptoms better in all cases	N/A
Teive et al, 2012	5	Jaw opening OMD secondary to Wilson's Disease	Botulinum toxin type A	Dystonia score partially reduced 3 weeks after injection	Mild dysphagia
Bhattacharyya et al, 2001	23	-5 with OMD -18 with Spasmodic Dysphonia	Botulinum Toxin	-Mean score +38.04 -OMD group derived smaller benefit (Glasgow Benefit Inventory Score)	N/A
Tan et al, 1999	162	OMD	-54.2±15.2 Units to masseter - 28.6±16.7Units to submentalis	-Mean global effect was 3.01 ±1 (4=complete abolition of dystonia)	-31.5% reported adverse effects -11.1% had dysphagia or dysarthria

Table 2.[7,8,9,10,11,12]

Case 1.
This patient was a 56 year old woman with a background of hypertension. She was diagnosed with a 3-4 year history of oromandibular dystonia characterized by increased mandibular movements affecting speech and mastication. The drug

history included prochlorperazine. Despite stopping this drug, symptoms persisted. Movements were a combination of jaw closing and jaw deflecting types. Over the course of a year, she received 3 courses of treatment with Botulinum toxin and is planned for ongoing treatment. After treatment, outcome measures show improvement in all categories.

Case 2.
This 45 year old diabetic woman presented with a history of chronic bilateral temporomandibular joint dislocations. Initial treatment with Botulinum toxin to the lateral pterygoid muscles arrested the dislocations but the patient developed a jaw retruding oromandibular dystonia. Treatment to the muscles of mastication resulted in alleviation of symptoms. The patient reported a negative overall benefit though she reported improvement in all categories. This case illustrates the difficulties in diagnosing OMD but it also show the versatility of Botulinum toxin treatment.

Case 3.
This 82 year old woman presented with OMD characterized by involuntary contractions of masseter, temporalis, digastric and the pterygoid muscles. She also had involuntary tongue movements. The treatment of the muscles of mastication was successful but the tongue movements remained an issue until the genioglossus muscle was treated. Because of her tendency for bruxism, the intraoral treatment of the lateral pterygoid muscles was difficult and an extraoral approach was used. The patient reported significant benefit from treatment. This was a mixed /combination type of OMD.

Case 4.
This 77 year old woman presented with OMD of the jaw deflecting type. She had a background of paroxysmal atrial fibrillation, hypertension and hypothyroidism. She was on warfarin treatment. The lateral pterygoid muscles were targeted for Botulism toxin treatment using an intraoral approach. The patient reported improvement in all categories.

Case 5.
This 57 year old man presented with a 4 year history of OMD with blepharospasm and was diagnosed with Meige's syndrome. He had a combination type of OMD characterized by jaw opening and jaw deflecting movements. Treatment was successful as reflected in the patient GBI score.

Case 6.
This 45 year old woman presented with OMD characterized by lateral and protrusive movements of the mandible. Therefore she was diagnosed with a mixed/combination type OMD. Patient admitted to a history of using mephedrone (M-CAT). The outcome measures show improvement in all categories.

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Case	Features	Treatment	GBI outcome
1	-Increased speed of involuntary posterior mandibular movements during mastication -Increased lisp during speech	-25U to both Masseter m. -25U to both Temporalis m.	Overall +55 General 92% Social 30% Physical 33%
2	-Chronic bilateral TMJ dislocation	-20U to both Masseter m. -20U to L. Pterygoid m. -10U to both Temporalis m.	Overall -11 General 57% Social 37% Physical 17%
3	-Bruxism -Movements of mandible and tongue	-25U to both L. Pterygoid m. -20U to both Genioglossus m.	Overall +63 General 93% Social 43% Physical 37%
4	-Lateral mandibular movements	-25U to both L. Pterygoid m.	Overall +72 General 98% Social 47% Physical 40%
5	-Meige Syndrome -Lateral mandible movements -Progressive opening -Blepharospasm -Upper right limb postural tremor	-25U to both L. Pterygoid m. -15U to both Ant. Digastric m.	Overall +22 General 72% Social 33% Physical 30%
6	-Lateral and protrusive mandibular movement	-25U to both L. Pterygoid m. -15U to both Masseter m. -10U to both Temporalis m.	Overall +39 General 81% Social 33% Physical 30%

Table 3.

Discussion.

The results from the study were in keeping with previously published outcomes. Therefore, our experience further demonstrates that Botulinum toxin type A is an effective treatment for OMD with minimal complications.
OMD is often misdiagnosed and medical treatment is sometimes ineffective. The use of Botulinum toxin has proven to be an effective and easy treatment with only a few complications reported. Botulinum toxin type A blocks the release of acetylcholine at the presynaptic junction, producing a transient weakening of the muscle activity without systemic effects. It also has the advantage of being used with electromyography (EMG) guidance which permits individual treatment plans with lower doses than with conventional treatment. Although, OMD can cause considerable functional and psychological disability to the patient, injection with Botulinum Toxin type A is a safe and simple treatment option which is highly effective compared with other therapies.
In our experience, injecting the Botulinum toxin into the lateral pterygoid muscle can prove difficult in OMD cases where there is a high frequency movement or a wide displacing movement of the mandible. Therefore, to overcome this, an

extraoral approach was used where the needle was directed through the sigmoid notch as illustrated in figure 1.
The cases highlighted have shown the successful treatment for the different types of OMD, including Meige's syndrome using Botulinum toxin type A. Also illustrated was the use of this treatment for chronic dislocation of the temporomandibular joints.
This case series outlines Botulinum toxin type A as a simple but effective treatment for all types of OMD. Its versatility lends itself to treatment of mostconditions involving muscular spasms and the resulting ataxia and dysfunction.

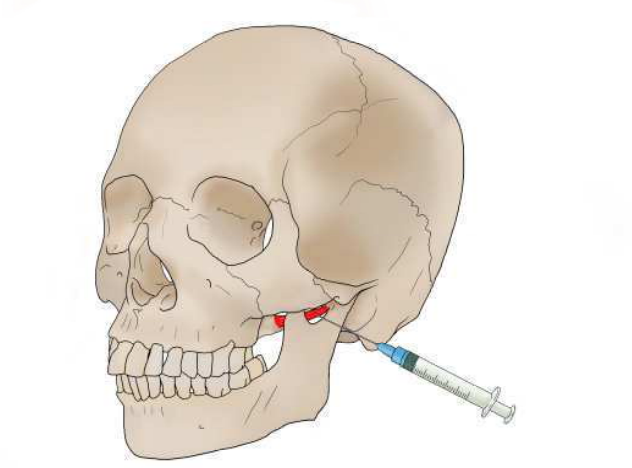

Figure 1.

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