Oromandibular dystonia: a case report of the lateral pterygoid muscle involvement and treatment with botulinum toxin A

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Abstract

The objective of the present case report is to punctuate the importance of individualized therapy procedures and the accurate diagnosis of the muscles involved in oromandibular dystonia and underline the role of electromyography (EMG). We report a woman who presented sustained jaw movement towards the left, severe difficulty in jaw opening and jaw protrusion. The patient was treated with injections of botulinum A toxin in lateral pterygoid, masseter, platysma, sternoclidomastoid, temporalis muscles with EMG guidance. She experienced an 80% reduction of her symptoms after the first injection. In jaw deviation dystonia symptoms impressively respond to botulinum toxin treatment of the pterygoid muscle. Individualized therapy procedures are necessitated.

Introduction

Oromandibular dystonia (OMD) is a form of focal dystonia, characterized by involuntary repetitive movements of the jaw and forceful contractions of the face.[1,2] The masticatory muscles are mainly affected however, lower facial and tongue muscles may also be involved.[3] It causes functional abnormalities in speech, swallowing and chewing and often leads to severe psychosocial discomfort. OMD is most often idiopathic. Botulinum toxin injection of the affected muscles remains the treatment of choice for OMD.[4,5]

Case Report

A 40-year old woman declared progressive speech difficulties, embarrassing grimacing movements and obstructed swallowing almost 2 years before the first visit to our out-patient clinic. At her admission she presented sustained jaw movement towards the left, severe difficulty in jaw opening during speech or eating, jaw protrusion and contraction of the platysma during speech and as a dystonic posture. She also exhibited mild bilateral exophthalmos. The severe dystonic movements and contractions were present most of the time forcing her to use external assistance to open her mouth whenever attempting to eat. The dystonic movements were only present during day time (Figure 1). The remaining neurological examination was normal.

Brain magnetic resonance imaging revealed no pathological findings. Levels of serum copper, ceruloplasmin and levels of urine copper were normal thus, dismissing Wilson's disease. Six months before her admission the patient underwent a complete thyroidectomy due to papillary thyroid carcinoma as biopsy revealed. Subsequently, the patient was placed on thyroid hormone replacement while, no subsequent improvement with regard to focal dystonia had occurred. Additionally, all the paraneoplasmatic antibodies were negative. For these reasons the paraneoplastic syndrome was ruled out. Finally, assiduous neuropsychological assessment was normal.

Oral medication including clonazepam, diazepam and anticholinergics proved to be ineffective. The patient was finally treated with injections of botulinum A toxin in the following muscles as shown on Table 1. To assess the effectiveness of this specific treatment video recordings were taken prior the treatment and one month post the toxin injection. For the evaluation of the improvement of the symptoms the dystonia movement scale (DS 0-8) was used.[6] We followed an extra-oral approach and therefore, the injection was conducted with electromyography (EMG) guidance using the Allergan's Electomyograph Signal Amplifier (Irvine, CA, USA). A needle electrode was inserted 3-4 cm until it was contacted with the muscle. Spontaneous lateral pterygoid muscle activity during rest exceeded guiding reference values. Hence, the dystonic activity of the muscle was confirmed.

One month after the bilateral treatment an obvious improvement both in speech and swallowing was observed. The patient herself experienced an 80% reduction of her prior symptoms while also no side effects were reported. The involuntary movements were abolished and she maintained correct occlusion without external assistance (Figure 2). The dystonia scale rating was remarkably reduced (Table 1).

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Table 1. Injected muscles treated with botulinum A toxin and dystonia movement scale (DS 0-8) during the first and the second treatment session.

	Dystonia movement scale		Injected muscles										Total units
	Mouth	Speech/ swallowing	Platysma		Lateral pterygoid		Temporalis		Masseter		Sternoclidomastoid		
			Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	
Baseline	4	2	-	-	-	-	-	-	-	-	-	-	-
1st injection	2	0	85		25	25	25	25	10	10	15		220
2nd injection	1	0	12.5	12.5	-	25	-	-	25	25	-		100

However, the difficulty in opening the mouth and the lateral jaw deviation persisted in a lesser degree. These were associated with the omission of injection of the upper head of the lateral pterygoid. Considering the reduced activity of the treated muscles and the alteration of the dystonic pattern the total units of injected toxin were minimized during the second treatment session. Botulinum A toxin was injected in the left lateral pterygoid muscle in multiple sites. One month later no mandibular displacement was observed and the patient experienced almost complete relief of the previous symptoms (Table 1).

Discussion

The dystonic features demonstrate a great variety in OMD. Therefore, the treatment has to be individualized in order to relief patients' symptoms. For practical reasons and based on the clinical picture, the jaw-dystonia has been classified as jaw-closing, jaw opening, or jaw-deviation dystonia.[3,7] Patients who suffer from jaw-closing and jaw-deviation dystonia have shown to have a better response to treatment when compared to those who have the jaw-opening subtype.[7] Most studies suggest botulinum toxin injection in contralateral lateral pterygoid and ipsilateral temporalis in jaw-deviation dystonia. Nevertheless, when jaw protrusion is combined, both lateral pterygoid muscles are treated. The muscles involved in jaw-closing dystonia include masseter, temporalis and medial pterygoid. Our patient is a rather complicated case which combines jaw-closing, jaw-deviation dystonia and jaw protrusion. Hence, the therapeutic protocol we followed was based to the EMG findings during injection. The recommended by the literature therapeutic process does not include platysma in the potential injected muscles. We additionally treated platysma with high doses of botulinum toxin A. This particular muscle depresses the mandible and soft tissue of the lower face resulting in tensing the skin of the neck.[8] Botulinum toxin treatment of the lateral pterygoid lead to a marked discomfort palliation which was estimated from the horizontal jaw movements' limitation for which this muscle is considered to be the most important.[9] Furthermore, the upper head of the lateral pterygoid muscle is mainly related to opening mouth movements while the lower head to the closing ones,[10] a reason for the upper head to be spared from botulinum A toxin treatment in a jaw closure case, as ours. A precise anatomic localization with the help of EMG guidance, the accurate injection of the lower head was achieved. The dystonic spontaneous activity of the muscle was found to be markedly decreased during the second injection.

Our patient presented a favorable response in the first treatment cycle. During the repeat treatment` the doses were markedly reduced (reduction by 40%), and the temporalis and sternocledomastoid muscles were omitted. Peak improvement, as shown by the scales was succeeded after the first botulinum toxin injection. The following treatment session did not demonstrate so striking results (Table 1), while the patient's satisfaction was increased, as there was a better cosmetic result. During the third and fourth follow up visits (12 and 16 months respectively after the initial treatment) the obsessive symptoms were abolished and subsequent injections were made only in the lateral head of lateral pterygoid muscle.

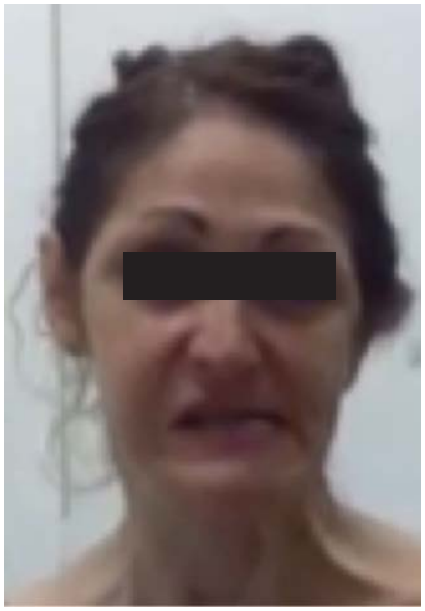

Figure 1. Mandibular displacement at baseline.

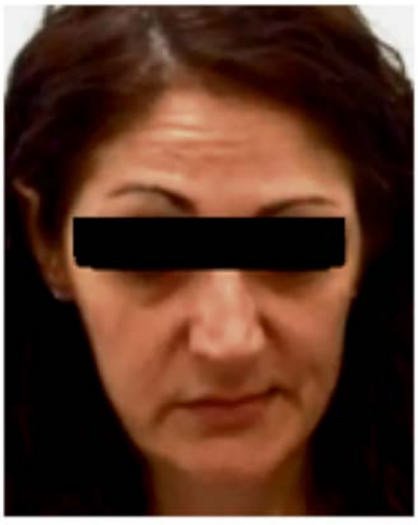

Figure 2. Mandibular displacement after first botulinum injection.

Conclusions

The jaw deviation dystonia is an uncomfortable neurological condition resisting to oral medication. Symptoms impressively respond to botulinum toxin treatment of the pterygoid muscle. Individualized therapy procedures are necessitated as the clinical manifestations greatly vary in OMD. Therefore, the accurate diagnosis of the muscles involved is of a great importance in order to accommodate effective treatment.

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