

Case Report

Jaw-Closing Oromandibular Dystonia Induced by Speaking in a Patient with Systemic Sclerosis

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Abstract

Oromandibular dystonia is a rare form of focal dystonia caused by involuntary spasms of masticatory, lingual and pharyngeal muscles. Here we describe a 53-year-old edentulous woman with systemic sclerosis who presented with dysarthria due to bilateral contraction of her masseter muscles during speaking. An anticholinergic medication was prescribed instead of botulinum toxin injection. Her condition markedly improved after medication. We suspected that edentulosity caused an impairment of proprioception in the oral cavity leading to subsequent development of dystonia.

Key words: Dystonia; Raynaud's phenomenon; Peri-implantitis

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Introduction

Dystonia is a neurological movement disorder wherein sustained muscle contractions cause twisting and repetitive movements or abnormal postures. The disorder may be hereditary or may be caused by other factors such as birth-related or other physical trauma, infection, poisoning etc.

Oromandibular dystonia (OMD) is a focal dystonia involving the masticatory muscles, muscles of facial expression and those of the tongue and pharynx. Involuntary, inappropriate, repetitive, or sustained muscle contractions cause varying degrees of jaw opening, closing, deviation, protrusion, or retrusion as well as facial grimacing, abnormal tongue or pharyngeal movement, or any combination of these.^{2,3} Because the actions of eating and speaking activate the dystonia, these tasks are particularly affected.⁴ Jaw dystonia induced by speaking is very rare.⁵ Here we describe a case of oromandibular dystonia in an edentulous woman with systemic sclerosis.

Case report

A 53-year-old female primary school teacher and a known case of systemic sclerosis presented to the Department of Rheumatology, Bangabandhu Sheikh Mujib Medical University with difficulty in speech for 6 months. She was diagnosed as a case of systemic sclerosis on the basis of tightening and thickening of the skin of the face, Raynaud's phenomenon and skin biopsy findings in 2008. She was prescribed tablet methotrexate 10 mg weekly with folate supplement, tablet prednisolone 5 mg daily and sustained release tablet nifedipine 10 mg twice daily. Her condition gradually improved thereafter and she continued those medications.

She developed difficulty in speaking in the form of variable slurring of speech six months back which gradually worsened over time. There was no specific diurnal variation of the problem. There was no history of difficulty in chewing and swallowing and limb weakness. She has history of incurring facial injury

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in 1994 while being a victim of violence and all of her teeth had to be extracted subsequently. She was treated with dental implants, but she ceased using dental implants as she suffered from recurrent peri-implantitis.

After being admitted in Bangabandhu Sheikh Mujib Medical University, we found her asymptomatic during resting phase of her oromandibular muscles (Fig 1), but on initiation of speaking there was spasm of her oromandibular muscles (Fig 2).

Examination of the patient revealed edentulosity, mild skin thickening in the periorbital area, microstomia and dysarthria. Partial improvement of dysarthria was demonstrable after putting a candy inside her mouth. Nervous system examination was otherwise normal. Both temporomandibular joints were normal. MRI of brain was normal. A diagnosis of jaw-closing dystonia was made.

She was prescribed tetrabenazine initially at a dose of 12.5 mg once daily, which was continued for a week. Dose was increased by 12.5 mg every five days up to 75 mg twice daily, which resulted in complete resolution of the dystonia. The drug was maintained at that dose thereafter.

Discussion

Oromandibular dystonia (OMD) refers to involuntary spasms of masticatory, lingual and pharyngeal muscles. Phenomenologically, there are six types of OMD: jaw-closing dystonia (JCD), jaw-opening dystonia (JOD), jaw-deviation dystonia (JDD), lip and perioral dystonia, lingual dystonia, pharyngeal dystonia, and combination OMD. OMD may be seen in isolation (focal dystonia) as part of a more widespread segmental cranial dystonia, or as part of a multisegmental or generalized dystonia.⁶

Focal OMD is rare. The prevalence of focal OMD varies, reportedly as high as 6.9 cases per 100,000. Women seem to be affected more frequently than men, with the onset typically between the age of 45 and 70 years.⁷

The pathophysiology of dystonia is unclear, but it is thought to originate in the centrally mediated dysregulation of movement due to defect in the basal ganglia, particularly in the sensory motor regions of the putamen. The mechanism of peripherally induced dystonia is also based on the theory of sensory pathway disruption at the level of the basal ganglia.⁸⁻¹⁰



Fig 1. Resting position



Fig 2. Masseteric spasm during speaking

OMD can be idiopathic, either focal or as part of generalized dystonia, or secondary to medications, trauma, metabolic disorders or orodental factors (e.g., edentulosity, ill-fitting dentures).¹¹⁻¹⁴ The patient described here had edentulosity and history of facial injury.

OMD typically causes involuntary jaw-opening or jaw-closure, tongue protrusion, dysarthria, and dysphagia.⁴ Initially dystonic episodes may be triggered by specific tasks such as eating, speaking or swallowing.¹⁴⁻¹⁵ Jaw dystonia induced by speaking is very rare.⁵ Later less specific motor tasks induce the symptoms and in advanced stages dystonic movements can occur at rest.¹⁴⁻¹⁵ Sensory tricks in oromandibular dystonia include touching the face or inserting something such as candy or the tip of a pencil into the mouth.⁴ The patient mentioned here had jaw-closing variety of OMD triggered by speaking and partial improvement was demonstrable after putting a candy inside her mouth.

The diagnosis of adult-onset primary focal or segmental dystonia is made clinically. Neuroimaging studies are useful if an underlying cause is suspected. But in most cases findings are generally normal. Simultaneous EMG recording of agonist and antagonist muscles may show inappropriate co-contraction, but this is not required for diagnosis.⁴

OMD is difficult to manage and its treatment has been limited to minimizing the symptoms of the disorder. There are several treatment options that can relieve some of the symptoms of dystonia, so physicians can select a therapeutic approach based on each individual's symptoms.¹⁷⁻¹⁹ Treatment approaches used to manage OMD include medication, BTX, local anesthetic blocks, dental appliances, behavioral modification and psychological support, and surgical procedures.¹⁷⁻¹⁸ Oral medication is the usual first line of treatment, but there is no medication to prevent dystonia or slow its progression. Tetrabenazine, clonazepam, or other oral drugs have been assessed in a systematic way in large studies. The results of oral medication for OMD have been largely disappointing. Some authors find the oral medication baclofen to be useful for OMD.¹⁸⁻²⁰

Botulinum toxin (BTX) injection into the affected muscle with or without EMG guidance is a second line therapy.^{8,18} BTX has been proven to be superior to medical treatment, particularly in focal dystonias.²¹ Injections of small amounts of this chemical into the affected muscles prevent muscle contractions, and they can provide temporary improvement in abnormal postures and movements characterizing dystonia.⁸ The toxin decreases muscle spasms by blocking the release of the neurotransmitter acetylcholine which normally causes muscles to contract. The effect is typically seen a few days after the injections, and it can last for several months before the injections need to be repeated.^{2,8}

Physical therapy, use of splints, stress management, and biofeedback may also help individuals with certain forms of dystonia.^{17,18} Peripheral denervation or myectomy is seldom needed since OMD usually responds well to BTX and must be delayed while other treatment options are effective.^{17,18}

Conclusion

Because of its rare occurrence, patients with OMDs are probably often misdiagnosed, or their diagnosis may be delayed. Consequently, these patients may also receive incorrect treatment, and the symptoms may worsen over the years. This case gives us the lesson that OMD should be born in mind while evaluating a patient with dysarthria.

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