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OROMANDIBULAR DYSTONIA: A RARE ENTITY

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ABSTRACT

Oromandibular Dystonia (OMD) is a rare neuromuscular disorder characterized by involuntary repetitive muscular contraction affecting different parts of the oromandibular region. It is an extremely debilitating disease and can have significant impact on the physical and psychosocial wellbeing of the patient but is often misdiagnosed due to lack of established diagnostic and management criteria. This review paper aims to highlight useful information regarding the clinical features, diagnosis and various therapeutic options for OMD. All the practitioners should be aware of the protean features of this disease so that it can be timely diagnosed and managed.

INTRODUCTION

The term dystonia was coined by Oppenheim in 1911.[1]Dystonia is a hyperkinetic movement disorder characterized by sustained or intermittent involuntary muscle contractions that cause abnormal, often repetitive, movements, postures, or both.[2,3]It may show a wide variety in its clinical presentation (varying from barely noticeable to severely disabling) depending upon the musculature affected, severity and distribution. [4, 5]

Dystonia is considered as part of the spectrum of dyskinesia and can have a profound effect on the personal, professional, and social life of a patient even leaving him/her unfit to live independently.[6] It can be classified on the basis of (a) aetiology(b) age at onset of symptoms (c) distribution of body regions affected.(Table I)[3]

Like other body parts, the head and neck regions are also affected by dystonia which are termed oromandibulardystonias. Hence, oromandibular dystonia (OMD) is a rare neuromuscular disorder characterized by involuntary repetitive muscular contraction affecting different parts of the oromandibular region[2].

OMD can befocal or part of a generalized dystonia. It can either be primary or secondary to medications, trauma, metabolic disorders or other neurologic movement disorders.[7,8,9] On the basis of anatomic location, it can be classified as 1) Jaw opening Dystonia 2) Jaw closing Dystonia. 3) Lip and Perioral Dystonia 4) Lingual Dystonia 5) Laryngeal Dystonia and 6) Combination Dystonia.[5,10]

OMD is usually seen in older adults, has a female predeliction and may follow a long term history of movement disorder.[6.] It generally involves masticatory muscles, muscles of facial expression, and those of the tongue and pharynx.

The clinical features in OMD usually depend upon the muscle or group of muscles involved which may include 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.[5]

In jaw opening dystonia, there is sustained contraction of the lateral pterygoid muscle which results ininability to close the mouth. Prolonged jaw opening may lead to difficulty in mastication, swallowing, speech and cause drooling[11]. Often idiopathic jaw opening dystonias aremisdiagnosed as dental problems, bruxism or temporo-mandibular joint disorders.[12,13] Jaw closing dystonia can occur alone or in association with jaw opening dystonias. The affected muscle is masseter which results in sustained trismus and jaw clenching. It is a task specific dystonia and is generally found in musicians who play wind instruments.[14]

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Table I Classification of Dystonia[3]

I.	On the basis of Aetiology	
1	Primary (or idiopathic)	Dystonia is the only clinical sign and there is no identifiable exogenous cause or other inherited or degenerative disease. Eg: DYT-1 dystonia
2	Dystonia plus:	Dystonia is a prominent sign, but is associated with another movement disorder. There is no evidence of neurodegeneration. Eg: Myoclonus-dystonia (DYT-11)
3	Heredo-degenerative	Dystonia is a prominent sign, amongst other neurological features, of a heredo-degenerative disorders.Eg: Wilson's disease.
4	Secondary	Dystonia is a symptom of an identified neurological condition, such as a focal brain lesion, exposure to drugs or chemicals Eg: Dystonia due to a brain tumour, off-period dystonia in Parkinson's disease
5	Paroxysmal:	Dystonia occurs in brief episodes with normalcy in between. These disorders are classified as idiopathic (often familial although sporadic cases also occur) and symptomatic due to a variety of causes.
II.	**On the Basis of Age at Onset**	
1	Early onset (variably defined as 20–30 years)	It usually starts in a leg or arm and frequently progresses to involve other limbs and the trunk.
2	Late onset:	It usually starts in the neck (including the larynx), the cranial muscles or one arm. Tends to remain localized with restricted progression to adjacent muscles
III.	**On the basis of Distribution**	
1	Generalized	Both legs and at least one other body region (usually one or both arms)
2	Focal	Single body region (e.g. oromandibular dystonia, writer's cramp, blepharospasm)
3	Segmental	Contiguous body regions (e.g. cranial and cervical, cervical and upper limb)
4	Multifocal	Non-contiguous body regions (e.g. upper and lower limb, cranial and upper limb)
5	Hemidystonia	Half of the body (usually secondary to a structural lesion in the contralateral basal ganglia)

Lingual dystonia affects the intrinsic muscles of the tongue. Primary or idiopathic lingual dystonia is very rare and therefore it is important to evaluate for secondary causes including medications, head injury, electrical injury, varicella infection etc.[15] In a large series of cases, Esper *et al* reported that 41% of cases of lingual dystonia were secondary to medications, 18% heredodegenerative and post encephalitic, 12% generalized dystonia and 29% focal primary lingual dystonia.[13,16] Lingual Dystonia is also a characteristic of pantothenate kinase associated neurodegeneration (PKAN), Lesch-Nyhan syndrome Wilsons disease and Meige Syndrome.[17] The clinical features of lingual dystonia include repetitive to sustained tongue tip protrusion or contraction which can be action induced with speaking, eating and whistling. In can affect speech and swallowing .In severe cases it is associated with tongue bitingand has even caused life threatening airway obstruction.[18,19]

Laryngeal or spasmodic dystonia is an action induced dystonia that affects the laryngeal muscles. Laryngeal dystonia can be classified as adductor type or abductor type. The adductor type accounts for 80% of laryngeal dystonia and causes spasms of vocal fold adductor muscles resulting in inappropriate closing of the vocal folds with speech. In this type, extreme effort is exerted to achieve fluent speech and the patient's voice quality is harsh and strained with voice breaks. The abductor type is rarer, with uncontrolled spasms of the vocal fold abductors resulting in speech with sustained breathiness and breathy voice breaks, sometimes to the point of aphonia. Mixed laryngeal dystonia has characteristics both of the adductor and abductor types.[13]

Management

Evaluation of a patient with OMD requires a complete history especially a detailed drug history. It is essential to exclude all secondary causes like drugs, trauma, infections, associated syndromes etc.

A full neurological examination followed by an MRI must be performed to evaluate for stroke or mass involving the basal ganglia. A blood creatinine kinase, ceruloplasmin level, and slit lamp exam to rule out Wilson's disease should be done. Routine lab tests are usually normal in these patients.[4] Also a thorough evaluation of the temporomandibular joint should be performed to rule out nonreducing TMJ disorders which can sometimes mimic jaw closing dystonia.[5]

Therapeutic options for OMD include systemic medications, botuliniumtoxin(BTX) injections, local anaesthetic blocks, speechtherapy and the use of oral sensory devices.

The first line therapy for dystonias is often medication. Anticholinergic drugs act by centrally inhibiting the parasympathic system thus reducing muscle spasm. Benzodiazepine decreases monosynaptic and polysynaptic reflexes by increasing presynaptic GABA inhibition. Anticonvulsants such as carbamazepine reduce severe muscle spasm by decreasing polysynaptic response. Carbidopa/levodopa in low dose may help dopa-responsive dystonia.[5,20]Effectiveness of medical therapy for OMD is variable in the literature but only approximately 17% of patients with OMD responded reported significant benefit from medical therapy.[8]

Botulinium toxin has been statistically proven to be superior to medical therapy, specially in focal dystonias.Injections of BTX-A into muscles of the base of the mouth, muscles of mastication and muscles of tongue have been found to show improvement in the symptoms but sometimes Injecting into the muscles can be difficult, as a very precise dose needs to be given to avoid weakening the muscle too much and some muscles are difficult to inject. Because of these difficulties muscles are usually injected using electromyography (EMG).BTX can however cause certain side effects like jaw weakness, loss of smile, dysphagia and nasal regurgitation which can be minimized through dose adjustment and improved technique.[5]

Sensory tricks can be used as an adjuvant to pharmacologic therapy to temporarily reduce the symptoms. For example, gently touching the lips or chin, chewing gum, talking or placing a finger near an eye or underneath the chinor humming in cases of laryngeal dystonias.

"Gesteantagoniste" or oral sensory feedback devices can also be used as an adjunct therapy. The use of an oral sensory device has been shown to decrease the frequency and dose of BoNT required to treat OMD.Eg; For jaw opening dystonia, thedevice is a custom molded retainer that fits the mandibular teeth. Over the molars there is an extra prominence that, when the patient bites down, stimulates the lateral pterygoid muscle to overcome the dystonic action and results in relaxation of the muscle.[13]

The special needs of dystonia patients should also be taken care of including social, emotional & nutritional problems. In cases of depression or social isolation, a psychological intervention must be seeked while a dietetic referral is required when dysphagia is present. The dietitian can suggest appropriate use of texture modified diets, food fortification and nutritional supplement drinks In cases where speech is affected, referral should be made to a speech and language therapist.[21]

CONCLUSION

Oromandibular dystonia is a very rare disorder with a wide spectrum of clinical presentation. The diagnosis of OMD is purely clinical and there is no gold standard to confirm the diagnosis. As a result, it is often misdiagnosed and subsequently patients are managed incorrectly.

Hence it is important that the private practioners have a thorough knowledge of dystonias, so that a timely diagnosis can be done by through a detailed case history, complete examination and multiple investigations to rule out other causes.and prompt management is done. BXT is the most accepted treatment for dystonias.

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