

Two cases of oromandibular dystonia referred as temporomandibular joint disorder

A. Viswanath and S.M. Gordon

University of Maryland Dental School, Baltimore, MD, USA

Corresponding address: Dr Sharon M. Gordon, University of Maryland Dental School, Baltimore, MD, 650 West Baltimore Street, Room 6255, Baltimore, MD 21201, USA.

Email: sgordon@umaryland.edu

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Abstract

Oromandibular dystonia is thought to originate from a dysregulation of centrally mediated movement, whereby repetitive or sustained spasms of the masticatory, facial, or lingual muscles result in involuntary, and possibly painful jaw movement. Here we present 2 unique cases of oromandibular dystonia referred as temporomandibular joint disorders.

Keywords

Temporomandibular joint disorders; Meige syndrome; oromandibular dystonia.

Case reports

Case 1

A 78-year-old white woman referred by her oral surgeon for evaluation of masticatory dysfunction presented on a mobility scooter with slurred speech and chief complaint of “jaw spasm, difficulty chewing, and change in bite” ongoing for 4 years. Dental history included mandibular left quadrant implant placement 4 years earlier, resulting in paresthesia of her lower lip secondary to surgical injury to the inferior alveolar nerve. She was subsequently treated by an oral surgeon who removed these implants 1 month after placement; this did not alter the sensory deficit. A year later, she began to experience facial spasms with mandibular deviation to the right. She had several repeated occlusal adjustments, but lost 2 teeth due to a fracture in the upper right quadrant reported as progressively worsening over the 4 years. She had been treated with low dose botulinum toxin A injection by her neurologist to differentially weaken the facial muscles on the left side.

Her medical history included seasonal allergies, asthma, moderate acid reflux, osteoporosis, and occasional vaso-vagal episodes since 2000. Her neurologic history included herniated disk C5–6 with radicular left arm pain, irritable bladder, past polio as a child resulting in left leg degeneration, and peripheral neuropathy with coldness and tingling in both feet.

Surgical history included tonsillectomy as a child, hysterectomy, removal of multiple dermal basal and squamous cell carcinomas and melanoma, hysterectomy, and surgical repair of a fractured right ankle.

Current medications included fexofenadine HCl and pseudoephedrine HCl, fluticasone with salmeterol inhaler, furosemide, amlodipine, omeprazole and ibandronate tablets monthly. Supplements included calcium carbonate, vitamin D weekly, glucosamine/chondroitin MSM daily and multivitamins C and E daily.

Her family history was significant for juvenile and adult onset diabetes mellitus, myocardial infarction, and throat and prostate cancer. She also reported a familial history of amyotrophic lateral sclerosis 4 (ALS 4). She was a non-smoker and denied oral parafunctional habits, but wore a night guard prescribed by her dentist for her symptoms. She stated that her stress levels increased 3 years ago due to her husband's health problems, and that her symptoms worsened at that time.

Cranial nerve examination and parasthesia mapping were performed as part of a head and neck examination. The cranial nerve examination was unremarkable. There was parasthesia in the right lower lip and vestibule, but not in the extraoral labia. The masticatory muscles were not tender to palpation; mandibular range of motion was a maximum vertical opening of 44mm, protrusive: 03mm, and left lateral excursion of 06mm. Right lateral excursion could not be measured because she could not move to the right. No joint sounds were detected by palpation or auscultation.

Differential diagnosis for unilateral involuntary movement of the jaw and face are reviewed elsewhere^[1,2]. Based on her history and absence of fasciculation or twitching, our differential diagnosis included ALS, polio or oromandibular dystonia. We referred her to a physical therapist in conjunction with continued botulinum toxin A injection by her neurologist and to a speech therapist. Discussion with her neurologist confirmed a diagnosis of secondary oromandibular dystonia, possibly related to ALS and exacerbated by the mandibular nerve injury.

Case 2

A 58-year-old white woman was referred for evaluation of craniofacial pain with chief complaints of daily headaches, bilateral ear fullness and bilateral temporomandibular joint sounds. She reported that routine activity aggravated the headache, relieved by over-the-counter Excedrin® Migraine. She stated that she sometimes experienced sharp pain on the right side of her head "like a pinched nerve", relieved by bending her head and raising the right shoulder. She reported facial pain, present upon awakening in the morning and persisting all day. She also reported hearing loss in the left ear and dizziness since a swimming accident as a teenager, left-sided tongue deviation, history of dysphagia with difficulty swallowing solids, and the sensation of "food being caught" in the throat. She reported excessive drooling, starting 2 years ago, and reported being told she had Meige syndrome/blepharospasm. The patient also reported difficulty walking after sitting, and pain in her feet and ankles. Palmar erythema was noted and she stated that her hands were becoming progressively weaker, with pain in both palms.

The patient's medical history was notable for heart failure diagnosed 6 years ago with ongoing tachycardia. In addition, she reported gastroparesis, controlled diabetes, controlled hypertension, hypothyroidism, steato-hepatitis, enlarged spleen and chronic obstructive pulmonary disease with obstructive sleep apnea. Her psychiatric history included dissociative disorder, major depression, obsessive compulsive disorder, anxiety with pain attacks, suicidal ideation, and multiple psychiatric hospitalizations, including a history of electroshock therapy. Surgical history included 2 left eardrum repairs at age 18 years, tonsillectomy, gall bladder removal and tubal ligation.

Her family history was significant for systemic lupus erythematosus and multiple sclerosis, but her antinuclear antibody (ANA) to screen for autoimmune disorders was negative. She was a current smoker, but denied substance abuse or alcohol use. She reported occasional jaw clenching while sleeping.

Medications included levothyroxine, Excedrin® Migraine, metoprolol succinate, lovastatin, fenofibrate, metformin ER, clonazepam, mirtazapine, bupropion, esomeprazole, trileptal and albuterol inhaler.

The cranial nerve examination was unremarkable. There was tongue deviation to the right with mandibular deviation to the left with opening, and drooping of the lower left lip at rest. Pain rating was 2 on a scale of 0 to 10 for facial pain at the time of the visit. Examination findings revealed mandibular range of motion with maximum opening of 54mm, protrusive excursion of

Table 1. Putative causes of oromandibular dystonia^[1,2,4]

Primary oromandibular dystonia	Neurodegenerative disorders
	Huntington disease
	Wilson disease
	Polio
	Amyotrophic lateral sclerosis (ALS)
Secondary oromandibular dystonia	Structural lesions and/or neural insults
	Lack of oxygen
	Inherited abnormalities
	Trauma
	Vascular
	Stroke
	Infectious
	Toxins and medications
	Heavy metal poisoning
	Dopamine receptor blocking agents
	Tricyclic antidepressants
	Selective serotonin reuptake inhibitors
	Lithium
	Antiparkinsonian medications
	Anticonvulsants
	Histamine H2 receptor blockers
	Autoimmune deficiency
	Metabolic
	Disorders of amino acid metabolism
	Disorders of lipid metabolism
	Disorders of mitochondria
	Lesch-Nyhan syndrome
	Neoplasms
	Psychological factors
	Stress
	Depression
	Fatigue
Psychogenic oromandibular dystonia	Psychological factors
	Stress
	Depression
	Fatigue

05 mm, left lateral excursion of 14 mm and right lateral excursion of 12 mm. She reported pain with these movements. There were no joint sounds detected by palpation and auscultation. Neck musculature was also painful to palpation and there was generalized body pain in areas palpated using body sites selected by the American College of Rheumatology for fibromyalgia examination.

Previous magnetic resonance imaging (MRI) reports showed left temporomandibular joint anterior disk displacement with reduction, and a normal disk position on the right with moderate degenerative changes in the right condyle. Brain MRI reports showed minimal microvascular ischemic changes in the cerebral white matter. Spinal MRI reports showed hypertrophy indicative of spinal stenosis.

Based on her history and physical presentation, the differential diagnosis included psychogenic facial spasm, tardive dyskinesia, or oromandibular dystonia^[1] with associated masticatory muscular pain and she was referred to a neurologist for a comprehensive diagnostic investigation.

Discussion

The terms oromandibular dystonia, craniocervical dystonia or Meige syndrome are often used to describe a focal or segmental dystonia whereby repetitive sustained spasms of the masticatory, facial, or lingual muscles result in involuntary, and possibly painful, jaw movements^[3,4].

Oromandibular dystonia is a rare condition; misdiagnosis is common and may be mistaken for temporomandibular joint disorders^[5-7] or other movement disorders^[2,3]. The diagnosis of dystonia is challenging because it is based on clinical findings that can be affected by several factors at the time of presentation as well as the psychological status of the patient and the training of the clinician^[3,8]. Oromandibular dystonia is characterized by reduced inhibition and abnormal plasticity at various motor system levels as well as the presence of sensory abnormalities. The mechanism is not well understood but is thought to originate from defects in the basal ganglia^[3]. However, dystonia is classified based on clinical characteristics and according to the affected muscles^[9].

As described in these 2 cases, the muscles involved may be muscles of mastication, leading to misdiagnosis as temporomandibular joint disorder, bruxism, or another dental problem^[7,10,11]. Patients may present with abnormal jaw opening such as jaw deflection, jaw retrusion or a combination^[7,12]. Dystonic spasms may also result in lip and tongue dyskinesias, and mouth retraction^[7,13]. Other associated symptoms may include eating dysfunction^[14,15], dysarthria, dysphagia, dysphonia, breathing difficulties and alteration in vocalization^[10], depending on the muscles involved. Table 1 describes putative causes of dystonia. Exacerbating factors such as stress, depression, fatigue and chewing are often reported, along with compensatory behaviors^[15]. Certain medications might also induce oromandibular dystonia as reviewed by Colosimo et al.^[16]

Treatment options for oromandibular dystonia are quite varied, as reviewed by Clark et al.^[12] Pharmacological therapy is usually the first line of management^[17] with botulinum toxin A injections currently the mainstay of treatment^[15,17]. Surgical therapies are generally a last resort for treating individuals with only certain types of dystonia^[17].

Teaching points

The differential diagnoses for unilateral involuntary movement of the jaw and face are many^[1,2] and can include temporomandibular joint disorder. It is important for health care providers to be familiar with oromandibular dystonia, as it can develop after dental treatment and is often misdiagnosed as a dental condition. Like patients with temporomandibular joint disorder, those with oromandibular dystonia may also present with accompanying psychiatric conditions such as depression, anxiety, obsessive compulsive disorder, and other psychological conditions, which may further confound the diagnosis.

References

1. Thompson PD, Obeso JA. Focal dystonia of the jaw and the differential diagnosis of unilateral jaw and masticatory spasm. *J Neurol Neurosurg Psychiatry* 1986; 49: 651-6. doi:10.1136/jnnp.49.6.651.
2. Tan NC, Chan LL, Tan EK. Hemifacial spasm and involuntary facial movements. *QJM* 2002; 95: 493-500. doi:10.1093/qjmed/95.8.493.
3. Lee KH. Oromandibular dystonia. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2007; 104: 491-6. doi:10.1016/j.tripleo.2007.04.001.
4. Albanese A, Barnes MP, Bhatia KP, et al. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. *Eur J Neurol* 2006; 13: 433-44. doi:10.1111/j.1468-1331.2006.01537.x.
5. Nutt JG, Muentner MD, Aronson A, Kurland LT, Melton III, LJ. Epidemiology of focal and generalized dystonia in Rochester, Minnesota. *Mov Disord* 1988; 3: 188-94. doi:10.1002/mds.870030302.
6. Clark GT, Stiles A, Lockerman LZ, Gross SG. A critical review of the use of botulinum toxin in orofacial pain disorders. *Dent Clin North Am* 2007; 51: 245-61, ix. doi:10.1016/j.cden.2006.09.003.
7. Balasubramaniam R, Rasmussen J, Carlson LW, Van Sickels JE, Okeson JP. Oromandibular dystonia revisited: a review and a unique case. *J Oral Maxillofac Surg* 2008; 66: 379-86. doi:10.1016/j.joms.2006.11.028.
8. Blanchet PJ, Rompre PH, Lavigne GJ, Lamarche C. Oral dyskinesia: a clinical overview. *Int J Prosthodont* 2005; 18: 10-19.
9. Albanese A. Dystonia: clinical approach. *Parkinsonism Relat Disord* 2007; 13(Suppl 3): S356-61. doi:10.1016/S1353-8020(08)70030-0.

10. Moller E, Bakke M, Dalager T, Werdelin LM. Oromandibular dystonia involving the lateral pterygoid muscles: four cases with different complexity. *Mov Disord* 2007; 22: 785-90. doi:10.1002/mds.21304.
11. Sankhla C, Lai EC, Jankovic J. Peripherally induced oromandibular dystonia. *J Neurol Neurosurg Psychiatry* 1998; 65: 722-8. doi:10.1136/jnnp.65.5.722.
12. Clark GT. Medical management of oral motor disorders: dystonia, dyskinesia and drug-induced dystonic extrapyramidal reactions. *J Calif Dent Assoc* 2006; 34: 657-67.
13. LeDoux MS. Meige syndrome: what's in a name? *Parkinsonism Relat Disord* 2009; 15: 483-9. doi:10.1016/j.parkreldis.2009.04.006.
14. Papapetropoulos S, Singer C. Eating dysfunction associated with oromandibular dystonia: clinical characteristics and treatment considerations. *Head Face Med* 2006; 2: 47. doi:10.1186/1746-160X-2-47.
15. Maestre-Ferrin L, Burguera JA, Penarrocha-Diago M, Penarrocha-Diago M. Oromandibular dystonia: a dental approach. *Med Oral Patol Oral Cir Bucal* 2010; 15: e25-7.
16. Colosimo C, Suppa A, Fabbrini G, Bologna M, Berardelli A. Craniocervical dystonia: clinical and pathophysiological features. *Eur J Neurol* 2010; 17(Suppl 1):15-21. doi:10.1111/j.1468-1331.2010.03045.x.
17. Goldman JG, Comella CL. Treatment of dystonia. *Clin Neuropharmacol* 2003; 26: 102-8. doi:10.1097/00002826-200303000-00010.