Oromandibular Dystonia—Meige’s Syndrome: Report of a Rare Case with Review

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ABSTRACT
Meige’s syndrome is a combination of two forms of dystonia; blepharospasm and oromandibular dystonia (OMD). Oromandibular dystonia (OMD) is a form of focal dystonia affecting head and neck region, including the lower face, jaw, tongue and larynx. It may be a factor contributing to muscle stiffness, degenerative changes in temporomandibular joint, mucosal lesions, damage to teeth, and dental prosthesis. We take this opportunity to present a patient with oromandibular dystonia associated with blepharospasm and spasmodic dysphonia.

Keywords: Meige’s syndrome, Oromandibular dystonia, Blepharospasm, Dysphonia.

INTRODUCTION
Oromandibular dystonia (OMD) is referred both as Meige’s syndrome and Brueghel’s syndrome.1 It has been alleged that Peter Brueghel, the Elder, illustrated the syndrome in 16th century in one of his paintings.2 Since the original description by Henry Meige of ten patients affected with orofacial dystonia which he called ‘spasm of facial median’, only a few reports of patients suffering from this disorder have been published in the literature.3 This condition is most commonly seen in middle aged or elderly with ratio of 2:1 female predominance.4 It is characterized by spontaneous, repetitive, nonrhythmic, symmetric dystonic spasms, first involving the orbicularis oculi muscle that begin unilateral, but soon becomes bilateral and progress to muscles of the lower face, jaw and tongue thereby making difficulty in speech and swallowing.5 The dyskinesia may fluctuate considerably from day to day, aggravated by emotional stress, fatigue and even in normal functioning like eating or talking. Idiopathic blepharospasm and oromandibular dystonia may occur separately or together. Usually thought to result either from an extrapyramidal disorder or as a complication of phenothiazine, it was also hypothesized that disruption of dental proprioception may be a contributing factor in progression of such disorder.6

CASE REPORT
A 60-year-old male patient reported to the Department of Oral Medicine and Radiology, with a complaint of difficulty in chewing, swallowing and speaking for the past one and a half year. While narrating the history, his speech was slurred and tongue was particularly dystonic. The patient had no adverse habit of tobacco use. Mental status examination of the patient showed no evidence of dementia. The patient gave the history of intake of muscle relaxant, which he was taking on and off on previous consultation with a local practitioner. Extraoral examination revealed vigorous and sustained grimacing of lower face region for every 30 seconds with spasm of jaw closure, mouth puckering, lip pursing and blepharospasm of both the eyes (Fig. 1). Moreover, a spasm of jaw opening, mouth retraction and difficulty in opening eyes, even for brief duration, was also evident (Fig. 2).

Contraction of perioral musculature and platysma worsened the spasms. At the time of examination the severity of spasms were recorded and tabulated (Table 1). On intraoral examination, partially edentulous state, periodontitis, pooling of saliva and lingual dystonia were also present.

Panoramic and sectional radiograph of the temporomandibular joint revealed degenerative changes in the condylar head region. Barium swallow was carried out for functional assessment in which bilateral valliculae, pyriform sinuses, esophagus, and gastro-esophageal junction were found to be delineated with contrast (Figs 3A and B).

Magnetic resonance imaging of the brain was performed. T1, T2W spin echo and IR images in axial, sagittal and coronal planes showed diffuse prominence of cerebral sulci, ventricular system and basal cisternae suggesting diffuse cerebral atrophy (Figs 4A and B). Imaging of cervical spine revealed normal lordotic curvature except for vertebral bodies that showed mild degenerative changes.

Based on the signs of spasmodic dysphonia, blepharospasm, repeated spasms of orofacial musculature and platysma, while jaw opening and closure, lip pursing, mouth retraction and specialized imaging investigations revealing cerebral atrophy, the diagnosis of orofacial dystonia (Meige’s syndrome) was made. On consultation with neurologist, the patient’s condition is being treated with baclofen, clonazepam, anticholinergic (trihexphenidyl) medications and multivitamin supplements.

DISCUSSION
Meige’s syndrome is a combination of two forms of dystonia; blepharospasm and oromandibular dystonia (OMD). The symptoms of blepharospasm are mainly uncontrollable closing of eyes, photophobia and closure of eyes during speech.
Oromandibular dystonia symptoms include spasms, jaw opening and closing, dysphagia and dysarthria. The tongue is usually involved as in lingual dystonia. Dystonic spasms may extend to involve the muscles of eyelids, nose and neck that are aggravated and worsened on mastication and speaking.

The cause of oromandibular dystonia is obscure. In adult form, without blepharospasm, mutation has been mapped to chromosome 14q22 in the GCH1 gene. Being aggravated by emotional stress, fatigue and depression, the proposed etiology could be attributed to defective inhibitory control of the basal ganglion of the forebrain, thalamus and medulla oblongata.

Oromandibular dystonia can be acute or delayed (tardive) adverse reaction to the administration of neuroleptics or prolonged use of antipsychotic medication. Meige’s syndrome is usually misdiagnosed as familial (inherited) cranial dystonia, that occurs in conjunction with idiopathic torsion dystonia, parkinsonism and even temporomandibular joint disorder as the patient exhibits task specific movements appearing only during activities, such as talking or chewing, which use the involved muscles, but it can progress to the point where the spasms are continuous. Diagnosis of Meige’s syndrome is made by proper history, and recording the distribution and severity of spasms at the time of examination at regular intervals. Surface electromyography may aid in recording...
Table 1: Distribution and severity of spasms at time of examination

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<th>Upper face</th>
<th>Lower face</th>
<th>Oral Muscles</th>
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- **Upper face**: BS indicates blepharospasm; EbE, eyebrow elevation; EbF, eyebrow frowning; and Nas, nasal muscle spasms.
- **Lower face**: LP indicates lip pursing; LT, lip tightening; MR, mouth retraction; and Pty, platysmal spasms.
- **Oral muscles**: JO indicates jaw opening; MC, mouth closure; Pal, palatal spasms; PhM, pharyngeal muscles; FM, floor of mouth; and Tg, tongue.
- **Others**: Tz indicates trapezius; ISM, inspiratory muscles; and Abm, abdomen.

+++: Severe spasm, ++: Moderate spasm, +: Mild spasm, –: No spasms of eye and facial muscles, precisely. Magnetic resonance imaging is not usually of any particular value but it is entirely proper to carry out specialized imaging investigation to rule out structural abnormalities.

Oromandibular dystonia has a variable nature, therefore making it difficult to predict the prognosis of this disorder. Mainstay of treatment is to control the symptoms of spasms, pain, disturbed posture and function. There is unfortunately no cure, but occasionally patients may show improvement with time. Medications such as anticholinergic, baclofen, and benzodiazepines may provide symptomatic relief in patients with oromandibular dystonia. Botulinum injections may help with blepharospasm and can be used to suppress dystonia but it has no cure.11 A multidisciplinary approach is advocated involving dentist, physical and occupational therapist, psychologist and neurologist for proper management and rehabilitation of such cases.

**CONCLUSION**

Meige’s syndrome is a dyskinesia of the eyelids and facial muscles. As to its fundamental cause, there is no much detailed information. Unfortunately, till today there is no permanent cure but current forms of therapy may provide the symptomatic relief to the patient.

The patient described in this case report is responding to medication and, on his subsequent follow-up, has little relief from the spasms and dystonia.

**ACKNOWLEDGMENTS**

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**REFERENCES**