CASE REPORT

Meiges Syndrome: An Unusual Cause of Involuntary Facial Movements - Two Cases with Literature Review

Soujanya Reddy[1], Gouramma Padanad[1]

ABSTRACT

Introduction: Meige's syndrome is a rare neurological movement disorder characterized by involuntary and often forceful contractions of the muscles of the jaw and tongue (oromandibular dystonia) and involuntary muscle spasms and contractions of the muscles around the eyes (blepharospasm).The cause of Meige syndrome is unknown. Researchers speculate that the cause of Meige syndrome may be multifactorial (e.g. caused by the interaction of certain genetic and environmental factors).

Case Report: It is a rare condition and only a limited number of cases have been reported in literature.However, many patients may remain undiscovered or misdiagnosed.Our aim is to introduce such two cases referred to our department with chief complaint of difficulty in chewing, speech, oromandibular dystonia and dysphasia.

Conclusion: Dental and allied health professionals are on occasion confronted with patients who exhibit abnormal facial movements. The specific symptoms and their severity vary from case to case. As an oral physician we should be well versed with this rare clinical presentation that are usually associated with serious underlying progressive neurologic disorder.Appropriate referral to the neurology service is essential and multidisciplinary approach is advocated involving dentist, physical, occupational therapist and psychologist for proper management and rehabilitation of such cases.

Keyword: Meige's syndrome, Oromandibular dystonia (OMD), blepharospasm, Botulinum

How to cite this article: Soujanya Reddy, Gouramma Padanad. Meiges syndrome: an unusual cause of involuntary facial movements - two cases with literature review. International Journal of Contemporary Medical Research 2015;2(5):1291-1293.

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Source of Support: Nil

Conflict of Interest: None

INTRODUCTION

Meige's syndrome is a type of dystonia.It is also known as Brueghel's syndrome and oral facial dystonia. It is actually a combination of two forms of dystonia; blepharospasm and oromandibular dystonia (OMD).[1] It has been alleged that Peter Brueghel the Elder, illustrated the syndrome in 16th century in one of his paintings.[2] The spontaneous occurrence of blepharospasm and dystonia movements in face muscles, particularly those of the perioral and mandibular regions, has been named as Meiges's disease which was first described by Henry Meigein 1910.[2]

It is characterized by spontaneous, repetitive, nonrythmic, symmetric dystonia spasms, first involving orbicularis oculi muscle that begin unilateral, but soon becomes bilateral and progress to muscle of the lower face, jaw, and tongue thereby making difficulty in speech and swallowing. It is not a psychiatric disorder.[3,4] The involuntary muscle contractions cause varying degrees of disability and pain, from mild to severe. The resulting involuntary movements and postures may significantly impact daily functional activities.[4]

The etiology of Meige's syndrome is uncertain. A disorder of basal ganglia function along with neurotransmitter imbalance (dopamine and acetylcholine) or complication of phenothiazine, is likely to be the mechanism involved in the causation of this disorder.It was also hypothesizes that disruption of dental propriception may be a contributing factor in progression of such disorder.[5]

The disease typically develops between 30 and 70 years of age and is more common in women.[3] The usual symptoms are irritation of eyes, blinking, and stiffness of the face. Approximately the incidence varies from 1 in 10,000 to 1 in 25,000.[6] It is a rare condition and only a limited number of cases have been reported in literature. However, many patients may remain undiscovered or misdiagnosed.

Our aim is to introduce two such cases of 57 years old female who were referred to our department with chief complaint of difficulty in chewing, speech, oromandibular dystonia and dysphasia and other patient of 62 year old male reported to the department with chief complaint of difficulty in mouth opening, chewing and speaking since 3 months. Both were further referred for treatment and management to neurological centre where they were treated successfully with Botulinum toxin.

CASE REPORT

Case 01

A 57 years old female patient reported to department of

oral medicine and radiology, with a complaint of difficulty in chewing, speech and repeated pursuing of her lips since 15years.

Patient gave history of repeated blinking of eyes, inability to open her mouth completely during speech since 6 months. Also gave history of spending her time in religious rituals which has been gradually increasing over period of time to the extent of more than 6hours daily, continuously chanting in front of god. She had a fear that not performing these rituals might harm her family members.

Patient had no deleterious habits. Past medical history, drug history, similar problem were negative. On general examination patient was cooperative, well oriented to time, place and person. Generalized weight loss was seen, speech was forceful with pooling of saliva with low pitched sound and occasional pauses. Vital signs were stable.

Extraoral examination revealed vigorous and sustained wincing movement of whole face, contraction of perioral musculature, persistent persuing of her lower lip with slurred speech and drooling of saliva and blepharospasm of both the eyes (figure 01, 02).Masticatory muscles were tender on palpation; mouth opening was normal measuring about 43mm from maxillary incisors to mandibular central incisors. No joint sounds were detected on palpation or auscultation.

Intraoral examination showed compromised periodontal status, indentation of teeth on lower labial mucosa & tongue suggested of morsicatio labialis & morsicatio linguarum due to persistent pursuing of lip &tongue(figure 02).

Clinical examination of motor nerve supply of face showed abnormal muscle movements of Frontalis (Unable to raise her eyebrows,loss of wrinkles on the forehead), Corrugator supercilii (Loss of nasolabial fold), Orbicularis (Abnormal twitching of eye bilaterally) &Buccinator(Opening and closing of jaw was abnormal)

Panoramic radiograph findings revealed no degenerative changes of condylar head. Based on clinical signsand history of contraction of perioral musculature, persistent pursuing of her lower lip with slurred speech, blepharospasm of both the eyes,orofacial dystonia with meige'ssyndrome (psychogenic) was given. Patient was referred to the neurocenter for the further treatment.

Case 2

A 62 year old male patient reported to the department with chief complaint of difficulty in mouth opening, chewing and speaking since 3 months.

On eliciting the history of the presenting illness patient gives history of fall from his bicycle, with direct impact on bridge of nose 3 months ago, following which he was conscious except for mild erosions and mild bleeding from nose. He was taken to nearby hospital for first aid and following which he started noticing difficulty in opening mouth, speech after four days. He also gave history of slurred speech, drooling of saliva and repeated blinking of eyes. Further history elicited from patient attender revealed while sleeping patient was opening his mouth wide unconsciously.

Medical and family history was non contributory. On general physical examination patient was poorly built poorly nourished well oriented to time and place. All vital signs and parameters were within the normal limit.Extra oral examination showed abnormal contraction of facial muscles,perioral and masticatory muscles.Mouth opening was restricted measuring approximately 12mm (figure 03).Temporomandibular joint movements were not appreciated due to restricted mouth opening.

Intraoral examination revealed partially edentulous with poor oral hygiene and compromised periodontal status with coated tongue. Subjective and objective symptoms of motor and sensory nerve appeared to be abnormal with respect to masseter, trapezius, sternocleidoid muscle.

Based on history and clinical findings provisional diagnosis of neuromuscular abnormality like Muscle dystrophy and Myasthenia gravis was given. To evaluate any presence of fracture of orofacial region further radiological investigation like panoramic and neck radiograph view were done and they did not reveal any fractures (figure 04). Other investigations like complete blood count – were within the normal limits. After the neurological examinations final diagnosis of orofacial dystonia with cervical spondylitis was given. Patient was educated and motivated for the physiotherapy and referred to the neurocenter for the further treatment.

DISCUSSION

Oromandibular dystonia (OMD) is a form of focal dystonia that involves masticatory, lower facial, labial, and lingual muscles.When OMD is combined with blepharospasmitis referred to as Meige's Syndrome named after Henri Meige,

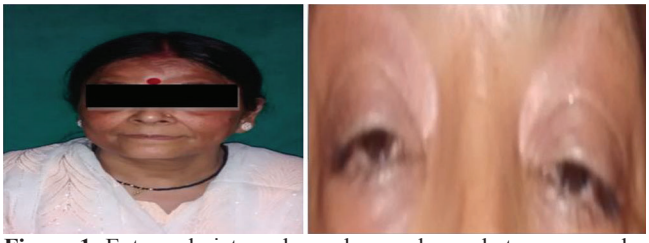

Figure-1: Extraoral picture showsabnormal muscle tone, muscular spasm & involuntry forcible closure of eyelids suggested of blepharospasm.

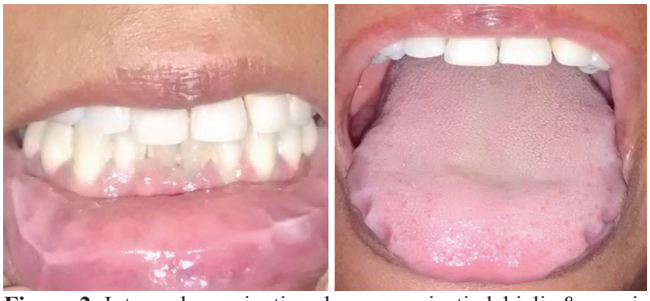

Figure-2: Intraoral examination shows morsicatio labialis & morsicatio linguarum due topersistent pursuing of lip & tongue

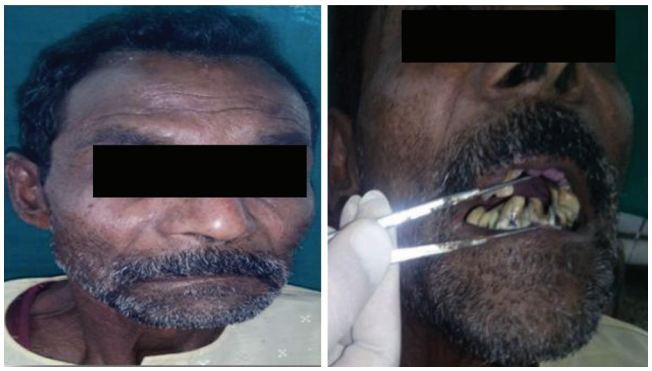

Figure-3: Clinical picture shows abnormal contraction of facial muscles & restricted mouth opening

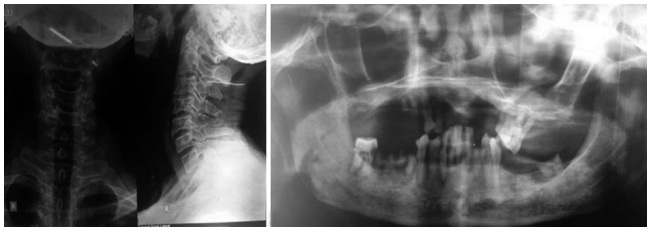

Figure-4: Neck radiograph shows cervical spondalytis andopg showsgeneralised periodontitis & no fracture evident.

the French neurologist who first described the symptoms in detail in 1910.[2]

The symptoms of blepharospasm are mainly uncontrolled closing of eyes, photophobia and closure of eyes during speech. The symptoms usually begin between the ages of 30 and 70 years old and appear to be more common in women than in men (2:1 ratio).[3] The etiology of Meige's syndrome is uncertain. Malfunctioning of a region of the brain known as the basal along with neurotransmitter imbalance (dopamine and acetylcholine) is likely to be the mechanism involved in the causation of this disorder. OMD can be acute or delayed (tardive) adverse reaction to the administration of neuroleptics or prolonged use of antipsychotic medication.Some cases of oromandibular dystonia occur in association with or secondary to another disorder such as tardive dyskinesia, Wilson disease, and Parkinson disease.[7]

The involvement of masticatory muscles in OMD may cause jaw-opening or –closing, lateral deviation, protrusion, retraction, or a combination. These movements often result in involuntary biting of the tongue, cheek, or lips and difficulty with speaking and chewing.

Furthermore, the lower facial muscles are often involved and may cause lip-pursing and grimacing, resulting in difficulty with pronunciation. In this presentseries first patient had similar findings of persistentpersuing of her lower lip with slurred speech and drooling of saliva and blepharospasm of both the eyes.

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.There is no way to detect Meige's by blood test or MRI or CT scans. OMD by itself may be misdiagnosed as TMJ.[8] OMD has a variable nature; therefore it is difficult to predict the prognosis of this disorder.

Treatment options for oromandibular dystonia are quite varied, as reviewed by Clark et al.[9] Pharmacological therapy is usually the first line of management with botulinum toxin. Surgical therapies are generally a last resort for treating individuals with only certain types of dystonia.[10]

Mainstay of treatment is to control the symptoms of spasms, pain, disturbed posture and function. There is unfortunately no cure, but is aimed at providing symptomatic relief to the patient.A multidisciplinary approach is advocated involving dentist, physical and occupational therapist, psychologist and neurologist for proper management and rehabilitation of such cases.

CONCLUSION

Meiges syndrome is a combined form of dystonia – involuntary, irregular muscle contractions – of the lower face, jaw and neck, as well as of the eye.There is no cure, although some patients may improve with time. Botulinum toxin injections may be used to suppress mouth spasms. Anti-spasm agents, muscle relaxants, or anti-convulsants have been reported to provide modest relief, at best, to some patients. Both the patients in this report responded to medication, provided symptomatic relief on subsequent follow up.

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