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## RESEARCH ARTICLE

### RARE NEUROLOGICAL DISORDER: MEIGE'S SYNDROME- A CASE REPORT

<sup>1</sup>Dr. Sagar Ramesh Ganvir and <sup>2</sup>Dr. Akash Sunjay Gupta

<sup>1</sup>MDS- III (ORAL AND MAXILLOFACIAL SURGERY), Post graduate student at Saraswati Dental College Lucknow

<sup>2</sup>Post graduate student at Saraswati Dental College Lucknow, 233, Faizabad Rd, Tiwari Ganj, Chinhath, Lucknow, Uttar Pradesh 227105

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#### ABSTRACT

Meige's syndrome is a rare neurological syndrome characterized by segmental oromandibular dystonia and blepharospasm. Its pathophysiology is not well known. A 45-year-old female presented to oral and maxillofacial department with blepharospasm and oromandibular dystonia with clinical provisional diagnosis of TMJ hypermobility. After thorough TMJ examination and physical examination including detailed neurological exam and psychiatric evaluation no TMJ disorder or any medical or psychiatric diagnosis could be made. The other differential diagnoses of extra pyramidal symptom, tardive dyskinesia, conversion disorder, anxiety disorder were ruled out by formal diagnostic criteria. accordingly with suspicion of Meige's syndrome she was referred to the neurologist and the diagnosis was confirmed. therefore, Meige's syndrome could be misdiagnosed as a TMJ hypermobility or psychiatric disorder such as conversion disorder or anxiety disorder because clinical features of Meige's syndrome are highly variable and affected by psychological factors and also can be inhibited voluntarily to some extent.

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#### INTRODUCTION

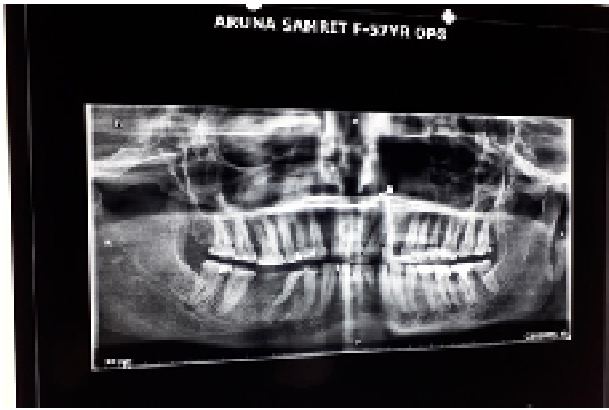
Meige's syndrome or Brughel syndrome are used by neurologists and other clinicians to describe the combination of blepharospasm and involuntary movements, forceful contraction of the muscles of jaw (oromandibular dystonia) and involuntary muscle spasm and contraction of muscles around eyes (blepharospasm). The disorder is named after Henri Meige in 1910, the French Neurologist who first described the syndrome (Debadatta, 2013). Its pathophysiology is not clearly known.

#### CASE REPORT

A 45-year-old female reported to the Department of oral & maxillofacial surgery with complaints of sideways deviation of the jaw since 3 years and grossly carious tooth. She visit to the local dentist for the same complaint resulted in this referral. At the beginning, the deviation was 3-4 times a day and that was not causing much difficulty for her. Symptoms frequency gradually increased and she also developed chin thrusting movement with also developed increased rate of blinking and

these movements were very embarrassing to her. With a progressive course the severity of symptoms increased causing difficulty in sleep and problem in daily routine activities. She could voluntarily inhibit the symptoms to some extent. The symptoms were precipitated with the emotional stressors, suggesting some secondary gain. No relevant past or family history of any medical or psychiatric illness was found. She was professionally chemist educated up to graduate. Mental status examination shows she is in anxious mood. There was no formal thought and perceptual disorder. She was well oriented, alert, and had normal fluency, repetition and naming. Her attention, calculation and abstraction were absolutely normal. General physical exam was unremarkable. Neurological exam showed intact cranial nerve, coordination, motor and sensory function normal. Ophthalmological exam showed no visual defect. Hematological examination are within normal limit—Haemoglobin, Differential Count, Total Leukocyte Count etc., were normal. Blood sugar, Liver Function Test, Thyroid function test, Computed Tomography Scan and Magnetic Resonance Imaging was within normal limit. Psychological assessment that is chemical and verbal abreaction could not reveal anything. The patient was sent to the neurologist for opinion and diagnosed as a case of Meige's syndrome. who advised investigations including radiographs, magnetic resonance imaging of the brain and right Temporomandibular joint.

\*Corresponding author: Dr. Sagar Ramesh Ganvir, MDS- III (ORAL AND MAXILLOFACIAL SURGERY), Post graduate student at Saraswati Dental College Lucknow.



The patient was kept on a trihexyphenidyl and Haloperidol 0.75 mg twice daily, also advised for extraction of grossly carious teeth. There was a little improvement on next follow-up.

## DISCUSSION

Dr. Horatio Wood, a Philadelphia neurologist, first drew attention to blepharospasm and other cranial dystonias in 1887<sup>(4)</sup>. Wood briefly mentioned facial and oromandibular dystonia in his textbook on disorders of the nervous system <sup>(4)</sup>.

The clinical features of this case suggestive of neurological disorder or conversion disorder. although points against diagnosis of conversion disorder were- long standing, presence of symptoms at difficult position, persistence of symptoms even at sleep. Since, there was no history of using any antipsychotic drugs, possibility of extrapyramidal symptom were ruled out. According to DSM IV TR criteria, anxiety disorder was also ruled out (Debadatta, 2013). Oromandibular dystonia and blepharospasm is referred as Meige's syndrome. Symptoms usually occurs between ages 30

and 70 year and more commonly affect woman than man (2:1) (Debadatta, 2013). Symptoms comprise of Oromandibular symptoms (difficulty opening mouth, grinding of teeth, spasm of jaw opening, sideways deviant of jaw, lip tightening) and Blepharospasm symptoms (increased rate of blinking, uncontrollable squinting, photophobia) (Akiyama, 1999). Sometimes dystonia may provoked by talking, chewing, biting applying pressure over chin. Generally involved muscles are masseter, temporalis and platysma (Ananth, 2000) The exact cause is not known, however, hypothesis of dopaminergic and cholinergic hyperactivity is most widely accepted. Current medical research says that the condition is caused by a defect in a large network of brain cells especially in basal ganglia. It is commonly misdiagnosed. Usually a neurologist specializing in movement disorder detect Meige's syndrome. No other definite test to detect the syndrome. Symptoms usually disappear with a sleep and present at activity, with some variation with a stress. There is no complete cure for Meige's syndrome (Poungvarin, 1997). Botulinum toxin injections in the facial musculature are considered the most effective treatment to date. Although the effects are temporary, the intramuscular injections provide rapid symptom relief (Jaffe, 1999).

## Conclusion

This case report emphasize the point that motor movement along with blepharospasm for long duration having definite stressors warrants and needs detailed evaluation. Meige's Syndrome, a rare neurological disorder should be considered as differential diagnosis of conversion disorder, like other neurological disorder presenting with Conversion symptoms like multiple sclerosis and myasthenia gravis.

## Footnotes

**Source of Support:** Nil.

**Conflict of Interest:** None.

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