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Review

MEIGE SYNDROME: A NARRATIVE REVIEW

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ABSTRACT

A particular combination of oromandibular dystonia combined with blepharospasm is known as Meige's syndrome. In order to avoid any misunderstandings, diagnosis is primarily clinical and necessitates a high index of suspicion. Striatal dopaminergic predominance is the most frequently accepted theory because the exact mechanism is unknown. Unfortunately, there is no cure, but injections of botulinum toxin and other medication classes, including anticholinergics, tetrabenazine, baclofen, and benzodiazepines, provide relief.

KEYWORDS: *Meige syndrome, spasm, dystonia, eye, muscle*

INTRODUCTION

Meige syndrome is one of the focused dystonic movement diseases, along with oromandibular dystonia and blepharospasm. Dystonia is characterized by abnormal, involuntary postures or motions of the body brought on by constant muscle contractions. Typically, neurological and medical issues are to blame. Meige syndrome comes under rare neurological movement disease characterized by uncontrollable, frequently startling contractions of the jaw and tongue muscles. French neurologist Henry Meige discovered aberrant midline facial muscle contractions in some patients in the early 20th century. He first referred to this as "spasm facial median." The clinical manifestation in the jaw and also oropharynx muscles of these patients was likewise comparable. In order to describe individuals with facial muscular spasms, notably blepharospasm and dystonia of the oromandibular muscles, Dr. George Paulson originally coined the name "Meige syndrome" in 1972 (1).

Meige's syndrome often manifests between the ages of 30 and 70. Women experience it twice as frequently as males. The typical symptoms include difficulty opening the mouth, tightness of the lips, jaw deviation, and teeth grinding. Some potential symptoms are blepharospasm, sensitivity to light, excessive squinting, and a higher blink rate. The masseter, temporalis, and platysma muscles are frequently involved. The actions of talking, chewing, and biting can exacerbate Meige's syndrome symptoms. Gum chewing, applying pressure to the chin, and sleeping help to relieve it. Numerous theories have been put up for its possible causes because the specific aetiology is unknown. The theory pointing to

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dopaminergic and cholinergic hyperactivity is the most frequently recognized, aside from idiopathic Meige's syndrome. The aetiology has also been linked to a defect in the basal ganglia region (2).

Typical and more recent atypical antipsychotics have been observed to cause acute and tardive Meige's syndrome, which promptly improves with antipsychotic withdrawal. Several pharmacological studies have proposed central dopaminergic predominance as a potential biochemical cause of Meige's illness. The finding further corroborates that it becomes better when dopamine-depleting medication tetrabenazine is used (3).

Meige's syndrome cannot be cured by any medical procedure. The most beneficial treatment to date is considered to be botulinum injections into the facial muscles. Other successful treatment approaches for Meige's syndrome include deep brain stimulation of the *globus pallidus internus* and electroconvulsive therapy. Only oral medications are utilised to treat most patients, and Meige's syndrome is often treated with various medications (4).

Epidemiology

The impacts of race and ethnicity have been implicated in the significant variation in incidence rates of segmental dystonia and blepharospasm. The epidemiology of primary dystonia from 14 chosen studies was examined by Defazio and colleagues. Segmental dystonia was present in 2-20% of people. Defazio et al. estimated that Segmental dystonia had a crude frequency of 59 per million (5).

In another Italian research focusing on 9 blepharospasm patients. The average age of onset for Meige's syndrome is in the sixth decade of life, and age seems to have been an independent risk factor in its development. Meige's syndrome affects more women than men, with a roughly 2:1 female-to-male ratio (6).

Marsden also noted that in all three forms of craniocervical dystonia, women were affected more frequently than men (7).

In research on 100 patients having blepharospasm and oro-facial dystonia, Jankovic and Orman showed a sizable woman:man ratio (women 62, men 38) (8). In addition, according to Duane (9), some oestrogen receptors that affect involuntary motor function put women at higher risk than men (10).

Pathophysiology

As previously mentioned, the most widely recognized theory for the pathophysiology of Meige syndrome is dopaminergic and cholinergic hyperactivity. However, it could also be brought on by the cortex's inhibitory neurons' (such as GABAergic neurons') diminished activity. Additionally, the patient is more susceptible to craniocervical dystonia due to a number of environmental and genetic variables. Positron emission tomography scans of the patients have revealed reduced blood flow towards the sensorimotor area in relation to lower facial vibrations, leading some researchers to hypothesize that these individuals have faulty sensorimotor functioning. In patients with Meige syndrome with isolated blepharospasm, silent "functional magnetic resonance imaging" has revealed reduced activation of the primary motor cortex, including the premotor cortex in the mouth region. It could be brought on by aberrant basal ganglia control over cranial nerve nuclei inside the brain stem. Individuals with craniocervical dystonia had reduced grey matter volume in the insular cortex, cerebellum, superior frontal gyrus, and calcarine fissure, as per brain imaging (11).

Additionally, genetics plays a role in the etiopathogenesis of focal dystonias. For example, the torsinA gene mutation seems to disrupt the flow of vesicles in and out of the nucleus at the cellular level. Dysregulation of transcription is the result of this. Other main focal dystonias are caused by a similar mechanism, such as CNS transcriptional factors (TAF1, THAP1). Studies using animal and human models provide evidence for the above-mentioned genetic abnormalities that may cause the neural network's growth to be altered (3).

Emotional stress may have a role in 33 percent of Meige syndrome patients, whose main symptoms include anxiety, sadness, and sleep issues (11).

Evaluation

It can be challenging to know where to begin the diagnostic process. For many, it starts at a primary care physician's office, an urgent care facility, or an emergency hospital. A diagnosis can become evident through these encounters, or it might take specialized testing and referrals. Dystonic anatomical classification and diagnosis continue to be clinical tasks greatly impacted by training and experience. Task-specific, subtle, and intermittent regional engagement is possible.

Additionally, many muscles have origins and insertions in many anatomical areas of the body. As a result, it may be challenging for many patients to distinguish between "focal" and "segmental" dystonia. For example, electromyographically-demonstrable minor participation of the upper thoracic paraspinous muscle is common in cervical dystonia patients having retrocollis (12).

Patients referred for neuropsychiatric evaluations and who had not responded to therapy in 49 cases were recorded over the course of multiple sessions. In the context of blepharospasm and oromandibular dystonia, Meige's syndrome was identified. The orbicularis oculi muscles' short, chronic, repetitive contractions and the facial, masticatory, labial, lingual, and mandibular muscles' dystonic involuntary motions were carefully observed. In addition, patients' medical histories and records were examined to weed out those who had taken neuroleptic medications. None of these individuals had ever used an antipsychotic drug, a neuroleptic, or anything else that could have caused parkinsonism or other uncontrollable movements. The results of the neurological and neuropsychological tests showed no intellectual, sensory, or pyramidal impairments. Similar negative results came from neuroradiological investigations, particularly brain computed tomography and magnetic resonance imaging (13).

Meeting with a doctor frequently during the diagnosis procedure may be beneficial and required. During the diagnostic phase, a primary care physician and specialist may suggest treatment alternatives to control symptoms. Doctors can also connect the patient with regional assistance programs, mental health services, and research opportunities.

Treatment

Meige syndrome is treated according to the distinctive symptoms that each patient shows. Botulinum A toxin (Botox) injections and medication therapy are the two main forms of treatment. After a injection, about 70% of people report improved chewing, swallowing, and spasm reduction (4).

Oral medicines are used to treat about a third of those affected (drug therapy). Unfortunately, the effects of these pharmacological therapies are typically mild or disappointing and frequently transient. No medication seems to work consistently well. Meige syndrome has been treated with clonazepam, trihexyphenidyl, diazepam, and baclofen (14).

The Food and Drug Administration has authorized the orphan drug botulinum A toxin as a therapeutic for blepharospasm, which has since become the main type of treatment. First, small doses of botulinum toxin are injected into the orbicularis oculi, paralyzing these muscles for many months. The process then has to be repeated. Many patients with blepharospasm have found relief from botulinum toxin injections, although not everyone does (4).

In rare circumstances, making particular motions, often referred to as "sensory tricks", may help people feel better. These motions include speaking, chewing gum, conversing, and lightly caressing the chin or lips. As a result, spasms may be reduced, the range of motion may be improved, and unaffected muscles may be strengthened through speech and swallowing treatment (15).

Various interventions fall within the categories of medicine and surgery as part of treatment. The effectiveness of anticholinergics, dopamine antagonists, and GABA receptor agonists for such individuals can be clearly understood once the process of Meige syndrome is understood. Antiepileptics, such as valproic acid, and a number of psychotropic pharmaceuticals are among the other medications. The particular GABA receptor complex components omega-1 and omega-2 are where eszopiclone and nitrazepam operate to reduce eyelid spasming. Some case studies claim that zolpidem is beneficial in treating these people because it is very selective for a GABA omega-1 receptor (16).

Eyelid spasms brought on by prolonged use of psychoactive substances can intensify through the use of olanzapine; however, they have been recorded with other antipsychotics as well.

Deep brain stimulation

In rare patients with Meige syndrome, deep brain stimulation (DBS) can result in a consistent, long-lasting relief of the symptoms of dystonia. DBS entails implanting a computerized pulse generator. The pulse is a thin metal electrode connected to generator as the brain's pacemaker. During office visits, a doctor programs settings that can be changed to best regulate symptoms. DBS functions by obstructing aberrant brain activity patterns (17).

Botulinum toxin and other conventional therapy approaches have not successfully treated patients when deep brain stimulation of the *globus pallidus interna* is used instead. The positioning of electrodes should be planned conceptually since the *globus pallidus interna's* ventral and posterior regions correspond topographically to the face, whereas the cervicofacial region is more anterior (18).

CONCLUSION

Meige syndrome is a rare spasm disease that must be taken in mind by clinicians, must be recognized, and has a different treatment protocol to be delivered.

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