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Meige Syndrome- A Case Report

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I. Introduction

Meige syndrome is a rare focal dystonic movement disorder characterized by blepharospasm (double eyelid spasms) and oromandibular dystonia. [1] Early and timely intervention in patients with Meige syndrome is essential for reducing morbidity and optimizing patient outcomes. Current treatment options for Meige syndrome include oral medication, botulinum toxin, DBS, and surgery in some cases. In the present report, authors present a case of Meige syndrome in a female.

II. Case Report

Patient was a 70 year old female who presented to her primary care clinic with a chief complaint of bilateral eyelids spontaneous spasms and jaw pains. She had been having these complaints for 3 months prior to her presentation, spasms were spontaneous in occurrence, repetitive in nature, and were associated with bilateral eyelid ptosis. She also complained of dryness and foreign body sensation in her eyes. She endorsed having to use her fingers to support her eyelids to keep them open. She also complained of constant blinking in both her eyes. She reported no improvement in her symptoms with over the counter eye lubricant drops. Her recent eye examination by her ophthalmologist was found to be normal. Patient had a past medical history of Type II Diabetes mellitus and was on metformin, primary open angle glaucoma, migraines, major depressive disorder, Grave's disease status post ablation, postablative hypothyroidism, hypertension, hyperlipidemia, GERD, asthma, overactive urinary bladder and diabetic neuropathy. On presentation, her vitals were within normal range. Her physical examination showed a Caucasian female, in moderate distress due to headaches and bilateral eyelids ptosis and spontaneous jaw twitchings. It was noted that she used her fingers to support her eyelids open during the examination. She was following up with a neurologist for her migraines as they had been getting worse in severity even with her current medication Ubrelvy 100mg. Her symptoms of lid ptosis and oromandibular pain were getting worse and were causing her trouble in her daily routine activities. She was started on botox injections for her migraines by her neurologist that seemed to wonderfully help her eyelid and oromandibular symptoms as well. She was then diagnosed as having Meige syndrome as a diagnosis of exclusion after the botox injections worked for her eyelid and jaw symptoms. At her most recent follow up, it was noted that her symptoms had completely resolved with the botox injections.

III. Discussion

Meige syndrome/ Brueghel syndrome/ idiopathic orofacial dystonia, is an unusual neurological disorder characterized by involuntary muscle contractions causing repetitive and stereotyped movements of the face, mouth, and neck.

Patients with Meige syndrome are typically aged between 30-70, with a mean age of 55.7 years. [2]

Although the exact pathophysiology of Meige syndrome is not known, it is hypothesized to be related to abnormalities in the basal ganglia. [3] Dystonia is believed to result from dysfunction in neurotransmitter systems, particularly affecting dopamine and gamma-aminobutyric acid (GABA). [4]

Primary Meige syndrome presents without an identifiable underlying cause and is typically characterized by isolated dystonic movements in the face and jaw. Meige syndrome can occasionally run in families, suggesting a hereditary susceptibility to the condition. Secondary Meige syndrome has been linked with identifiable triggers like psychological stress, exposure to certain drugs or toxins, or physical trauma such as surgery or a brain injury. There appears to be a tendency for the disorder to affect females more frequently.

As Meige syndrome cannot be diagnosed with a single specific test, a thorough evaluation is essential to exclude other potential causes and determine the severity of the condition.

Brain MRI plays an important role in ruling out structural abnormalities that may resemble or exacerbate dystonic symptoms, including tumors, vascular malformations, or other lesions.[6] Although brain MRI findings in Meige syndrome often appear normal, imaging is essential to distinguish secondary causes of dystonia. Additionally, MRI and/or computed tomography (CT) of the brain can help in ruling out stroke as a potential underlying cause.

Electrophysiological investigations are essential in the differential diagnosis of Meige syndrome. Nerve conduction studies and electromyography (EMG) can provide important information regarding muscle activation patterns and help eliminate other neuromuscular conditions that can be mistaken for dystonia.[6] Surface EMG recordings are particularly helpful for assessing the type and intensity of myofascial contractions in individuals with Meige syndrome.

Genetic testing is a thoughtful consideration in cases where there is a family history of dystonia or suspected genetic predisposition.[7] Genes linked to dystonia, such as *TOR1A (DYT1)*, *THAP1*, and *GNAL*, may be subjected to targeted sequencing or panel testing to help identify pathogenic mutations that may be responsible for Meige syndrome.[8]

Metabolic and toxicological screening is important in cases with unusual symptoms or suspected secondary dystonia. Laboratory testing for metabolic diseases, such as Wilson disease or mitochondrial abnormalities, can be considered in suspected cases. Additionally, investigating exposure to chemicals or drugs known to induce dystonia is suggested. The workup should include a serum drug screen, SSA/SSB levels, Cu and ceruloplasmin levels, uric acid levels, and Beck's Depression Inventory.[9]

Meige syndrome treatment focuses on abating the symptoms, improving functional outcomes, and enhancing the quality of life for affected individuals. Treatment approaches usually necessitate a combination of medications, botulinum toxin injections, surgical interventions, and supportive therapies.

Oral medications, including muscle relaxants, dopamine receptor antagonists, and anticholinergic drugs, are used to target the dystonic symptoms in Meige syndrome.[10] These medications focus on reducing muscle spasms and improving motor function.

Botulinum toxin injections represent an effective treatment modality for blepharospasm and facial dystonia. Affected muscles receive botulinum toxin type A injections that cause temporary chemical denervation and reduce muscular hyperactivity.[11]Treatment typically occurs every 2 to 6 months and is catered according to an individual's response and the degree of symptoms.[12]

Deep brain stimulation (DBS) is a consideration to be made for patients unresponsive to noninvasive treatments.[13] Electrode placement is meticulously planned, targeting the ventral and posterior segments of the globus pallidus interna, as these areas correspond to the facial region.[14]

Surgery can be an option for some cases unresponsive to medical or local therapy, aiming to enhance both functional and aesthetic outcomes. [15] Blepharoplasty correction, coupled with selective myectomy and myotomy, can provide lasting improvements for patients with refractory Meige syndrome.[16][17][18]

IV. Conclusion

Patients affected by Meige syndrome experience a wide array of complex symptoms, including involuntary blinking (blepharospasm) and spasms of the jaw muscles (oromandibular dystonia), which may progress eventually over time and significantly affect their quality of life. Management often includes a multidisciplinary approach, consisting of medications, botulinum toxin injections, and supportive therapies to alleviate symptoms and enhance function. Treatment challenges arise due to the disorder's unpredictable course and its impact on daily functioning, often affecting speech, swallowing, and chewing muscles.

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