Bosphorus Medical Journal
Boğaziçi Tıp Dergisi

Case Report

DOI: 10.14744/bmj.2024.53336

Bosphorus Med J 2024;11(2):57–59

Meige Syndrome (Blepharospasm with Orofasial Dystonia): Two Case Reports

Meige Sendromu (Orofasial Distoni ile Blefarospazm): İki Vaka Raporu

Hatice Ferhan Kömürcü

ABSTRACT

Meige syndrome is a rare movement disorder characterized by involuntary spasms of the muscles around the jaw, tongue, and eyes. It may manifest idiopathically or secondary to an underlying cause. Herein, two cases diagnosed with primary and secondary Meige syndrome are reported, and the literature regarding their clinical findings and treatment approaches is reviewed.

Keywords: Blepharospasm; Meige syndrome; Orofacial dystonia.

ÖZET

Meige sendromu, çene, dil ve göz çevresindeki kasların istemsiz kasılmaları ile karakterize, nadir görülen bir hareket bozukluğudur. İdiyopatik ya da altta yatan bir nedene sekonder olarak ortaya çıkabilir. Burada, primer ve sekonder Meige sendromu tanısı alan iki olgu rapor edilmiş olup; klinik bulguları ve tedavi yaklaşımları ile ilgili literatür gözden geçirilmiştir.

Anahtar sözcükler: Blefarospazm; Meige sendromu; Orofasiyal distoni.

Department of Neurology, University of Health Sciences Türkiye, Fatih Sultan Mehmet Training and Research, İstanbul, Türkiye

Cite this article as: Kömürcü HF. Meige Syndrome (Blepharospasm with Orofasial Dystonia): Two Case Reports. Bosphorus Med J 2024;11(2):57–59.

Received: 17.03.2024
Revision: 17.03.2024
Accepted: 02.04.2024

Correspondence:
Dr. Hatice Ferhan Kömürcü. Department of Neurology, University of Health Sciences Türkiye, Fatih Sultan Mehmet Training and Research, İstanbul, Türkiye

Phone:
+90 532 594 47 02

e-mail:
skomurcu@hotmail.com

Meige syndrome is a rare neurological movement disorder characterized by involuntary and often strong spasms of the jaw and tongue muscles (oromandibular dystonia) and involuntary spasms of the muscles around the eyes (blepharospasm). Meige syndrome is also known as "Brueghel Syndrome", "Idiopathic Blepharospasm-Oromandibular Dystonia Syndrome", "Segmental Cranial Dystonia", "Segmental Cranio-Cervical Dystonia", "Wood syndrome", "Blepharospasm plus" depending on different anatomical regions involved.[1]

Meige syndrome was first described by French neurologist Henry Meige in 1910.[1] The disease is more common in women than men. Different prevalence rates have been reported regarding blepharospasm and segmental dystonia.[1] The crude prevalence of segmental dystonia was reported by Defazio et al.[2] as 59 per million (95% CI 16-151). Symptoms generally start between the ages of 40-70 and can rarely be seen in younger individuals.[1]

The exact cause of Meige syndrome is unknown, and its primary or idiopathic forms are seen in most patients. Sometimes cases of secondary Meige syndrome develop due to long-term use of neuroleptics or underlying brain disorders. This syndrome has also been described in patients with essential tremor, Parkinson's disease and atypical Parkinsonism.[1]

Herein, two female patients diagnosed with primary and secondary Meige syndrome are reported.

Case Report

Case 1

A sixty-five-year-old female patient had complaints of involuntary movements and spasms of oromandibular and periorbital muscles that had been present for six years. It was learned that she had no history of head trauma, stroke, cerebral infection, rheumatological, malignant, or neurological disease. There were no obvious pathological findings in cranial images. Serum copper and ceruloplasmin levels were within their normal ranges. She did not describe any drug addiction or antipsychotic drug use. Her neurological examination was unremarkable except for oromandibular dyskinesia and blepharospasm. The patient was evaluated as compatible with primary Meige syndrome. Botulinum toxin was applied to the patient who did not accept constant medication use. The treatment was repeated at three-month intervals, and the patient benefited significantly from the treatment.

Case 2

A 60-year-old female patient was admitted to our was admitted to our clinic due to involuntary movements and spasms around her mouth and eyes, gradually increasing slowness in her movements, and reduction of spontaneous facial expression that had persisted for seven years. She had a history of long-term antidepressant and antipsychotic drug use. On neurological examination, perioral dyskinesia and blepharospasm, bilateral rigidity, walking with small steps in slight anteflexion, and hypomimia were noted. Her speaking rate had slowed down. There were no obvious pathological findings in cranial magnetic resonance imaging (MRI) examination. There was no history of rheumatological disease. Serum copper and ceruloplasmin levels were within normal limits. The patient was evaluated as compatible with secondary Meige syndrome and parkinsonism, which developed due to psychiatric drug use. The patient was advised to reduce the dosage of her psychiatric medications by the consultation of her primary doctor and botulinum toxin treatment was started. The treatment was repeated at three-month intervals. Perioral dyskinesia and blepharospasm symptoms significantly decreased with botulinum toxin treatment.

Discussion

There is no specific test to diagnose Meige syndrome. Diagnosis is made by a comprehensive clinical evaluation, a detailed patient history, and description of characteristic symptoms. We reported two cases of primary and secondary Meige syndrome. In our first case, no etiological cause that could cause Meige syndrome was detected. Our second case had a history of long-term use of psychiatric drugs. Both cases were female and their complaints started in their fifth decades.

Basal ganglia play a role in regulating motor and learning functions. Dysfunction in the basal ganglia of the brain may play a role in the development of Meige syndrome. Problems related to the basal ganglia in individuals with Meige syndrome are not fully known. Neurophysiological features are similar to focal dystonia, characterized by abnormal plasticity and impaired inhibition. The reason why Meige syndrome is common in women may be the tendency of estrogen receptors to increase involuntary muscle spasms.[1] Sometimes cases of oromandibular dystonia may be associated with or occur secondary to a disorder such as tardive dyskinesia, Wilson's disease, and Parkinson's disease.[1]

In most patients, Meige syndrome is primary or idiopathic. Genetic factors are important in the development of the disease in primary Meige syndrome. Patients with p.Gly213Ser or p.Ala353thr mutations have been found to have clinical findings of Meige syndrome.[1] It has been reported that GNAL (guanine nucleotide binding protein G, subunit alpha) mutations have a role in cranial and cervical dystonia, but further studies are required to clarify this issue.[3-6] In our first case, neurological examination findings were unremarkable except for oromandibular and periorbital dyskinesias and spasms. The patient had no other history of disease. Genetic analysis could not be performed for our patient. There were no obvious pathological findings on cranial MRI.

Our second case had a history of long-term psychiatric drug use and had signs of parkinsonism. There were no obvious pathological findings in blood tests and cranial MRI. Denervation hypersensitivity, thought to be due to central dopaminergic activity, may develop in one quarter of patients using neuroleptic drugs for more than one year. The denervation hypersensitivity hypothesis is the most widely accepted view in the development of secondary Meige syndrome.[6] There are changes in receptor functions that cause facial or cervical dystonia which improves with the use of dopamine-depleting agents. Drugs that increase central dopamine activity include antiemetics (metoclopramide), antipsychotics, antidepressants, selective serotonin reuptake inhibitors, antihistamines, and dopaminergic agonists. Additionally, Meige syndrome may develop in cases such as head trauma, stroke, demyelination of the brainstem region, normal pressure hydrocephalus, cerebral hypoxia, postop-

eratively (bilateral thalamotomy), kernicterus, space-occupying lesions, and encephalitis.[1,6] Meige syndrome may be accompanied by other movement disorders such as Parkinson's disease, Wilson's disease, olivopontocerebellar atrophy or Lewy body disease.[1,6-8]

In the differential diagnosis, spinocerebellar ataxia, progressive supranuclear palsy, Wilson's disease, ischemic stroke, autoimmune/inflammatory diseases (multiple sclerosis, lupus erythematosus, Behçet's disease), metabolic causes (hypoxia, pontine myelinolysis), neoplasms (meningioma, metastatic tumor), myoclonus-dystonia syndrome, generalized anxiety, and psychogenic craniocervical dystonia should be considered.[1,4]

In Meige syndrome, treatment is directed towards the symptoms. Anticholinergics such as trihexyphenidyl, GABA receptor agonists (diazepam and baclofen etc.), dopamine antagonists (tiapride and tetrabenazine etc.), antiepileptic drugs such as valproic acid, botulinum A toxin injections or surgical treatments including deep brain stimulation can be used in its treatment.[8-10] Drug treatment is effective in approximately 1/3 of patients; however, drug treatments are generally moderately effective and often provide temporary benefit.[8-10] Botulinum A toxin is used to treat muscle spasms associated with blepharospasm and oromandibular dystonia. Sometimes patients may experience relief of symptoms by sensory tricks which include biting a toothpick, chewing, speaking, or lightly touching the lips or chin.[1] Speech and swallowing treatment can reduce spasms, improve range of motion, and strengthen unaffected muscles.[1] Rarely, spontaneous remissions may also occur. Deep brain stimulation is used in the treatment of selected cases with Meige syndrome.[1] More research is needed to determine the long-term safety and effectiveness of alternative treatment modalities for Meige syndrome.

Meige syndrome is a rarely seen segmental dystonia characterized by involuntary spasms of the muscles around the jaw, tongue, and eyes. Primary (or idiopathic) and secondary Meige syndrome forms may occur. If a secondary cause is detected in patients, it is necessary to treat it accordingly. Botulinum toxin application is an effective treatment method in symptomatic treatment.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Use of AI for Writing Assistance: Not declared.

References

1. Pandey S, Sharma S. Meige's syndrome: History, epidemiology, clinical features, pathogenesis and treatment. J Neurol Sci 2017;372:162–70.
2. Defazio G, Martino D, Aniello MS, Masi G, Abbruzzese G, Lamberti S, et al. A family study on primary blepharospasm. J Neurol Neurosurg Psychiatry 2006;77:252–4.
3. Pedrero-Escalas MF, García-López I, Santiago-Pérez S, Vivancos F, Gavilán J. Clinical experience with patients with spasmodic dysphonia and primary Meige syndrome. Acta Otorrinolaringol Esp (Engl Ed) 2019;70:1–5.
4. Jochim A, Li Y, Gora-Stahlberg G, Mantel T, Berndt M, Castrop F, et al. Altered functional connectivity in blepharospasm/orofacial dystonia. Brain Behav 2017;8:e00894.
5. Horisawa S, Ochiai T, Goto S, Nakajima T, Takeda N, Kawamata T, et al. Long-term outcome of pallidal stimulation for Meige syndrome. J Neurosurg 2018;130:84–9.
6. Gautam P, Bhatia MS, Kaur J, Rathi A. Meige's syndrome. Ind Psychiatry J 2016;25:232–3.
7. Uemura T, Mochida M, Matsumi T, Yoshimoto K, Tatebayashi Y. Therapy-refractory schizophrenia in a patient who previously suffered from Meige's syndrome. Neuropsychopharmacol Rep 2019;39:297–300.
8. Jinnah HA, Factor SA. Diagnosis and treatment of dystonia. Neurol Clin 2015;33:77–100.
9. Karp BI. Botulinum toxin physiology in focal hand and cranial dystonia. Toxins (Basel) 2012;4:1404–14.
10. Sobstyl M, Ząbek M. Deep brain stimulation in the treatment of torticollis and Meige syndrome. Neurol Neurochir Pol [Article in Polish] 2011;45:590–9.