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"I Can't Open My Eyes": A Case of Blepharospasm and Apraxia of Eyelid Opening

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Chief Complaint: Difficulty Opening Eyes

History of Present Illness: A 72-year-old man presented to the oculoplastic clinic reporting an increasingly frequent difficulty opening his eyes for the past two years. He described being unable to open his eyes voluntarily, sometimes thrusting his head backwards or rubbing his brow with his fingers during these episodes. He was also bothered by frequent contractions of the muscles around the eyes on both sides of his face which caused forcible eyelid closure. He believes his inability to control eyelid opening was responsible for a driving accident a few months prior.

Past Ocular History: The patient suffered from bilateral ocular surface irritation, worse upon waking, for which he used preservative-free artificial tears.

Medical History: Previously repaired hip fracture after a motor vehicle accident.

Medications: None.

Family History: None.

Social History: Lives with his wife in a two-bedroom apartment. No smoking or alcohol consumption.

Physical Exam

- **Visual Acuity** (with correction): 20/25 Right eye (OD) and Left eye (OS)

- **Extraocular Motility:** Full both eyes (OU)
- **Pupils:** 4 mm dark, 2 mm light OD; 4 mm dark, 2 mm light OS, no RAPD OU
- **Intraocular Pressure:** 15 mmHg OU
- **Confrontation Visual Fields:** Full OU
- **Hertel Exophthalmometry:** 14 mm OU
- **External Examination:** Bilateral brow ptosis and dermatochalasis. Frequent spasms of the orbicularis oculi muscles, procerus, corrugators bilaterally, causing forcible eyelid closure. The patient had other episodes where he was unable to open his eyes voluntarily even in the absence of obvious contractions of the protractor muscles; during these moments, he would rub his temples or brows or thrust his head backwards. The upper eyelids were easily everted.
- **Slit Lamp Examination:** Mild inferior superficial punctate erosions and conjunctival hyperemia OU
- **Dilated Funduscopy Exam:** Normal disc, macula, vessels, and periphery OU

Course

The patient had evidence of blepharospasm with concurrent apraxia of eyelid opening (ALO). He also demonstrated brow ptosis, floppy eyelids, and dermatochalasis which were likely worsened by blepharospasm. The patient received 5 unit injections of botulinum toxin A into the procerus, corrugator, and at the medial and lateral junctions of the pretarsal and preseptal orbicularis in the upper lids. Injections of botulinum toxin offered symptomatic improvement and an obvious reduction in both the blepharospasm and ALO, although he required increased doses of botulinum toxin at subsequent follow-up appointments. When botulinum toxin failed to produce adequate functional improvements, the patient received bilateral upper eyelid blepharoplasty, pentagonal wedge resection for floppy eyelids, and limited myectomy of both upper lids. Three years later, he underwent bilateral direct browplasty.

He was lost to follow up and presented several years later after sustaining injuries from a motor vehicle collision which he attributed to his reduced visual function from blepharospasm and ALO (Video). He underwent additional injections of botulinum toxin A in the pretarsal orbicularis and glabella. One month after the injections he noted significant improvement in his visual function with decreased blepharospasm and apraxia of lid opening.

Video

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Discussion

Blepharospasm is characterized by bilateral, uncontrolled, involuntary spasms of the eyelid protractor muscles and brows, sometimes triggered by stress, intense light, or fatigue. Contractions of the procerus, corrugator, and orbicularis oculi are readily observed on clinical examination with depression of the brow (Charcot sign). During spasms, patients are unable to open their eyes. However, once obvious contractions cease, patients are able to readily initiate eyelid opening. Blepharospasm may occur independently or in association with other disorders of the orofacial muscles (Meige's Syndrome) or cervical muscles (Brüeghel's Syndrome). While the etiology is unclear, associations with essential tremor and Parkinson's disease suggest that blepharospasm may arise from dysfunction of the basal ganglia, although lesions at other cortical and subcortical structures have been identified as well.[1, 2] Because of the coexistence of ocular surface disease, lubrication with artificial tears and blepharitis management with warm compresses and eyelid scrubs should be considered prior to instituting more invasive modalities. FL-41 rose-tinted lenses have been shown to improve discomfort from photophobia as well as reduce blink rate and eyelid contraction force in patients with blepharospasm.[3] Blepharospasm is exquisitely sensitive to botulinum toxin injection into the eyelid protractors and this is often administered every three months. True failures of botulinum are rare, occurring in fewer than 2% of patients.[4, 5]

Apraxia of eyelid lid opening (ALO) is a condition which may occur concurrently with blepharospasm, or rarely, as an independent condition. Blepharospasm and ALO are frequently observed together in patients with advanced Parkinson's disease (PD) and Progressive Supranuclear Palsy (PSP). A separate review on the diverse ophthalmological features of PD can be found on EyeRounds (206-I-cannot-read.htm)

ALO is characterized by the intermittent inability to open the eyelids after closure in the absence of apparent contraction of the orbicularis oculi muscle. Unlike blepharospasm, where visible contractions of the eyelid protractors are easily witnessed, patients with ALO exhibit contractions of the frontalis muscle which elevates the brow, employ motor tics such as backward thrusting of the head, or palpate the periocular skin to encourage eyelid opening. ALO is not a true eyelid apraxia; it is better considered a focal eyelid dystonia because the patient's motor system is temporarily prevented from contracting despite normal understanding of the command.[6-8]

Two mechanisms are thought to be at work in ALO. The first is prolonged, involuntary pretarsal orbicularis contraction, where there is persistence of tone in the pretarsal orbicularis muscle despite a command to open the eyelids. The levator palpebrae superioris is unable to overcome its antagonist muscle and eyelid opening is prevented. The second mechanism involves involuntary levator palpebrae inhibition, where initiation of levator contraction is delayed after the command to open the eyelid is initiated.[6, 8, 9]

Botulinum injections into the orbicularis muscle have been largely successful in the management of ALO. Krack and Marion treated 34 patients with ALO, either isolated or coexistent with blepharospasm, PD, or PSP with approximately 30 units of onabotulinumtoxin A per side and found improvements in 83% of patients in all groups.[8] They also suggested that the junction of the pretarsal and preseptal portions of the orbicularis is a more efficacious site of injection, favoring it over injections in the orbital component. Boghen *et al.* found that lid metrics, such as lid opening latency and lid movement durations were prolonged in patients with ALO and improved by 30-38% in patients who were treated with botulinum injections.[10] In a review on the role of botulinum toxin, Jankovic *et al.* summarized the strong evidence for the use of botulinum toxin into the pretarsal orbicularis muscle for patients with ALO, particularly if coexistent with blepharospasm.[11]

The role of systemic dopaminergic therapy as another treatment for ALO has been suggested in select case reports. Lee *et al.* described a patient with PD who developed ALO after the dose of levodopa/carbidopa was reduced.[12] When therapy was resumed, the ALO resolved. Yamada *et al.* described a patient with PSP whose ALO was resistant to injections of higher doses of botulinum toxin, but responded to an augmented dose of levodopa/benserazide.[13] The mechanism at play is not well understood, but it is likely that supplementary dopamine at the level of the basal ganglia may improve the component of ALO resulting from involuntary inhibition of the levator muscle.

Deep brain stimulation of the subthalamic nucleus has been used as a therapeutic modality for PD and has been shown to induce ALO in some patients while improving pre-existing ALO in others.[14-16] The varied impact of subthalamic stimulation on ALO has not yet been explained, but it may relate to creating a lesion of unique structural and functional components within the subthalamic nucleus whose role on ALO is not yet understood. Another possibility is that electrical current from deep brain stimulation spreads to adjacent structures which impacts ALO.

In patients whose blepharospasm does not respond satisfactorily to botulinum toxin, the possibility of unwitnessed ALO must be considered. As mentioned, true failures of botulinum toxin are exceedingly rare, and it has been suggested that most patients whose blepharospasm does not respond to botulinum toxin have coexistent ALO. Rana *et al.* described two patients with ALO and blepharospasm and PSP who responded to an initial treatment with botulinum toxin but later stopped responding satisfactorily despite increased doses. Only after a partial orbicularis myectomy was performed in one, and eyelid crutches instituted in the other, did persistent botulinum toxin injections offer therapeutic benefit.[17] An approach which combines treatment modalities with botulinum toxin may be considered in patients with ALO who are refractory to botulinum alone.

Anderson *et al.* have been strong proponents for the role of surgical myectomy in patients with ALO coexistent with blepharospasm who are refractory to botulinum toxin.[5, 18, 19] They conducted a retrospective chart review of patients in whom blepharospasm was refractory to botulinum injections. Patients underwent a full orbicularis myectomy of the upper eyelids with removal of every filament of orbicularis muscle as well as removal of the procerus, corrugator, depressor supraciliaris, repair of associated eyelid malpositions, as well as punctal occlusion. Forty-five of the 51 patients had concurrent ALO, and 33% of patients had a completed "cure" of ALO, while 50% experienced some improvement in ALO symptoms. Complications from the surgery included orbital hemorrhage, post-operative ocular surface disease, forehead numbness, and the need for further surgery. This study was retrospective and subject to recall bias; moreover, the surgical procedure was not standardized as surgery for eyelid malposition and/or punctal occlusion was offered simultaneously. Despite these weaknesses, the study suggests a role for surgical myectomy in improving ALO refractory to botulinum toxin.[5]

Selected case reports have identified a potential role for brow lifting in patients who are refractory to botulinum and surgical myectomy. However, compromise of an already tenuous ocular surface from brow elevation must be strongly weighed against the potential benefit on ALO symptoms in patients with PD.[20]

Blepharospasm, characterized by bilateral involuntary contractions of the eyelid protractors, is exquisitely sensitive to injections of botulinum toxin. True failures of botulinum toxin are rare, and ALO must be suspected in these cases. ALO, better described as an eyelid dystonia, is often responsive to botulinum toxin injections into the pretarsal orbicularis muscle or at the junction of the pretarsal and preseptal orbicularis muscle. In patients who are refractory to botulinum toxin injections, other complementary therapies, such as eyelid crutches and surgical myectomy should be considered. Patients suffering from PD may benefit from augmented systemic dopaminergic therapy in consultation with the patient's neurologist. The role of more invasive treatments, such as deep brain stimulation of the subthalamic nucleus, remains to be elucidated. Further research which seeks to

quantify the electromyographic components of ALO will allow ophthalmologists to better understand the mechanisms at play in an individual patient's ALO and offer targeted treatments to improve patients' visual function and quality of life.

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