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Floppy Eyelid Syndrome

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Two cases illustrating the presenting symptoms and comorbidities associated with floppy eyelid syndrome

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PATIENT #1 INITIAL PRESENTATION

Chief Complaint

Eye irritation

History of Present Illness

The patient is a 42-year-old male who presents with a six week history of right eye irritation and discharge. He states that he has been experiencing burning, tearing, and a gritty feeling in his eyes nearly every day for the last six weeks and intermittently for several months before that. He notices that the symptoms are worse in the morning and are occasionally associated with a clear discharge from his eyes. Initially, he believed the symptoms were due to allergies, but the symptoms have persisted even as his nasal congestion and sneezing have resolved with over-the-counter allergy medications.

Ocular history

• Keratoconus both eyes (OU)

Medical History

Appendectomy

Medications

• 10 mg loratadine daily

Family History

- Father with hypertension
- Mother with history of migraine headaches

Social History

- Works in computer repair
- Drinks 1-2 beers on weekends
- No tobacco or drug use

Review of Systems

- Morning headaches
- Daytime sleepiness

OCULAR EXAMINATION

Visual Acuity

- Right eye (OD): 20/60, 20/30 with pinhole
- Left eye (OS): 20/40, 20/20 with pinhole

External exam

• OD: lateral ptosis, significant eversion of the upper eyelid with minimal upward traction; significant lower eyelid laxity



(../cases-i/case240/Fig1-LRG.jpg)



(../cases-i/case240/Fig2-LRG.jpg)



(../cases-i/case240/Fig3-LRG.jpg)

- OS: significant eversion of the upper eyelid with minimal upward traction
- External measurements:
 - o Palpebral fissure: 6.5mm OD, 8mm OS
 - o Margin reflex distance: 1.5mm OD, 2.5mm OS
 - o Levator function: 15mm OD, 15mm OS

Extraocular Motility

• Full OU

Pupils

• Equal, no relative afferent pupillary defect (RAPD)

Slit Lamp Exam

- Lids/Lashes: Lash ptosis OU
- Conjunctiva/Sclera: Bulbar conjunctival injection superiorly OD>OS; upper palpebral papillary conjunctival reaction OD.
- Cornea: Diffuse punctatr epithelial erosions (PEE) with central thinning OD, clear OS
- Anterior chamber: Deep and quiet OU
- Iris: Normal architecture OU

Lens: Clear OUVitreous: Clear OU

Dilated Fundus Exam

• Normal optic disc, macula, vasculature, and periphery OU

CLINICAL COURSE

Given the patient's nonspecific ocular complaints and an examination significant for right ptosis as well as right-sided papillary conjunctivitis and asymmetric corneal thinning, the diagnosis of floppy eyelid syndrome (FES) was suspected.

Due to the strong association between FES and obstructive sleep apnea (OSA), the patient was given the Epworth Sleepiness Scale screening form and returned with a score of 17/24, which indicated excessive sleepiness. At the conclusion of his first visit, the patient was referred to a sleep medicine specialist for polysomnography. Regarding symptom management, the patient was given an eye (Fox) shield for each eye and instructed to wear them while he slept until permanent options could be discussed at his next appointment. He was then scheduled for follow-up in clinic after completing his referral visits.

The patient returned for his follow up visit with the diagnosis of OSA. Since his OSA was now being treated with continuous positive airway pressure (CPAP), his ocular irritation and discharge had improved. He still noted tearing on the right, which was thought to be secondary to his eyelid laxity. Options for surgical correction of his FES versus continued observation were discussed. Due to the severity of his symptoms, the patient elected for surgical repair. An upper and lower lateral tarsal strip shortening procedure was performed bilaterally. A demonstration of this procedure (../video/plastics/6/4-floppy-eyelid-syndrome.htm) can be seen in this video.

PATIENT #2 INITIAL PRESENTATION

Chief Complaint

Right eyelid droop

History of Present Illness

The patient is a 66-year-old man brought to the clinic by his wife after she noticed that his right eyelid was drooping. She noticed his right eyelid droop recently but believes it has been this way for the past two years as evidenced by pictures in the family photo album. The patient denies irritation, pain, and discharge from both eyes. He states that he has had to tilt his head back when hunting in order to see the full visual field out of his right eye, which is his dominant eye.

Ocular History

• Myopia

Medical History

- Hypertension
- Hypercholesterolemia

Medications

- Simvastatin 40mg daily
- Lisinopril 20mg daily

Family History

- Father died of heart failure
- Mother died of lung cancer

Social History

- Retired schoolteacher
- Denies alcohol, drug, or tobacco use

PHYSICAL EXAM

Visual Acuity

- Right eye (OD): 20/20 with correction
- Left eye (OS): 20/20 with correction

External exam

• OD: Ptosis, significant eversion with gentle traction on the upper eyelid



(../cases-i/case240/Fig4-LRG.jpg)



(../cases-i/case240/Fig5-LRG.jpg)

• OS: Minimal eversion with gentle traction on the upper eyelid

Extraocular Motility

• Full OU

Pupils

• Equal, no RAPD

External

• Palpebral fissure: 5mm OD, 7.5mm OS

• Margin reflex distance: 0mm OD, 2.5mm OS

• Levator function: 15mm OD, 15mm OS

Slit Lamp Exam

Lids/Lashes: Ptosis OD; lash ptosis OUConjunctiva/Sclera: Clear and quiet

• Cornea: Clear OU

• Anterior chamber: Deep and quiet OU

• Iris: Normal architecture OU

Lens: Clear OUVitreous: Clear OU

Dilated Fundus Exam

• Normal optic disc, macula, vasculature, and periphery OU

CLINICAL COURSE

The patient's examination and history were consistent with FES. The patient's subsequent score of 20/24 on the Epworth Sleepiness Scale increased the clinical suspicion of FES in the setting of OSA. While the patient did not complain of symptoms, it is important to evaluate the tension of the eyelids in any patient undergoing possible eyelid surgery. A referral was made to a sleep specialist and a follow-up was scheduled.

The patient was subsequently diagnosed with OSA and returned to clinic two months after beginning treatment with CPAP. Although the patient's chief complaint was ptosis, it was explained to him that correction of his eyelid laxity would need to be performed prior to any ptosis surgery. The patient underwent an upper and lower lateral tarsal strip on the right. Four months later, the patient underwent ptosis correction of the right upper eyelid.

Discussion

Floppy eyelid syndrome (FES) is an ophthalmologic disorder characterized by laxity of the upper eyelids leading to spontaneous eversion of the eyelids during sleep and other activities. It is generally associated with several systemic, non-ophthalmologic conditions, such as obstructive sleep apnea (OSA) and obesity, and causes significant ophthalmic comorbidities of the conjunctiva, cornea, lid, and tear film.[1]

Prevalence

FES was originally described in middle-aged, obese men with papillary conjunctivitis in the setting of elastic, pliable upper eyelids. [2] While it is still most commonly diagnosed in overweight men between the ages of 40-69 years old, the condition has also been reported in a broader patient population, including women and children. [3,4] The diagnosis FES is frequently found in patients with comorbid conditions such as obesity, hypertension, diabetes, hyperlipidemia, hyperglycinemia, Down syndrome, and conditions causing chronic eye rubbing (e.g. psoriasis, cocaine use, epibulbar nodular fasciitis, etc.). [1,5]

One of the strongest comorbid associations in FES is with OSA, which may have both diagnostic and therapeutic implications. An association between the two conditions was first described by Gonnering and Sonelland in 1987.[6] Several studies have described the prevalence of OSA in patients suffering from FES and have found

that between 31.3% and 96% of patients with FES also have OSA.[5,7] Furthermore, a systematic review detailing the inverse relationship (the prevalence of FES in patients with OSA) found that up to 45.2% of patients with OSA suffered from FES[8]. The importance of mandatory sleep testing for any patient with floppy eyelid syndrome cannot be overstated. Because obesity, male gender, and age are all independent risk factors for both OSA and FES, the causality of the relationship is less clear. However, it is known that patients' sleeping postures, often affected by OSA, can determine the laterality of their symptoms. That is, the eye affected by FES typically corresponds to the side the patient sleeps on. If the patient alternates sides or sleeps face down, both eyes can be affected.[7] Furthermore, as the severity of OSA increases, as measured by the apnea-hypopnea index, the likelihood of FES increases.[9]

Patients with FES typically present with non-specific eye complaints that are often unilateral, but can be bilateral. Irritation, tearing, discharge, itching, foreign body sensation, and general discomfort of the eye may be mentioned.[5] The nonspecific nature of these complaints frequently leads to missed diagnosis, thus prolonging the patient's clinical course.

Pathophysiology

The pathophysiology of FES and the frequently encountered ocular comorbidities are primarily thought to be caused by mechanical irritation. The initial insult is presumed to be chronic mechanical injury from eversion and/or rubbing of the eyelid. Several authors have speculated these mechanical forces may lead to loss of tarsal elasticity from pressure-induced ischemia-reperfusion, abnormalities in elastin, enzymatic breakdown of elastin, abnormal eyelid apposition, genetic predisposition or some combination thereof.[10] Whatever the mechanism, the increased eyelid laxity results in the eyes becoming prone to exposure from nocturnal eversion and lagophthalmos. The prolonged and frequent nocturnal eversion results in exposure keratopathy that is nearly universally seen in these patients. Chronic irritation and inflammation of the cornea can progress to scarring and neovascularization. One of the most serious corneal complications associated with FES is keratoconus, which has been found in 4% to 32% of patients suffering from FES.[11] In addition, chronic irritation of the conjunctiva manifesting as papillary conjunctivitis is frequently seen, as well as meibomitis, eyelash ptosis, and loss of eyelash parallelism.[5] Table 1 summarizes the variety of ophthalmic pathology that may be associated with FES.

Table 1. Ocular findings associated with FES

Eyelid

- Dermatochalasis
- Blepharitis
- Blepharoptosis
- Meibomitis
- Upper lid lash ptosis
- Lower lid laxity
- Extropion (lower lid)
- Entropion (upper lid)

Cornea

- Superficial punctate kertopathy
- Erosions/ulcers
- Neovascularization
- Keratoconus

Sclera/Conjuctiva

- Papillary conjunctivitis
- Epidulbar nodular fasciitis
- Dry eye
- Pseudopterygium
- Keratoconjuncitivitis

Other

- Lipid tear film deficiency
- Glaucoma
- Meibomian gland dysfunction

Diagnosis

The diagnosis of FES is made clinically, defined broadly as rubbery, malleable, and easily everted upper eyelids. Many authors have attempted to more narrowly and precisely define the syndrome; diagnostic criteria have been proposed and include objective measurements of clinical findings such as horizontal distraction, snapback, and vertical lid pull.[5] lyengar et al. measured a series of patients' upper eyelid laxity and demonstrated that there was a statistically significant difference in the measurement of anterior eyelid distraction in the patients' symptomatic eyes vs. asymptomatic eyes.[12] Regardless of the specific criteria considered, the diagnosis of FES should be considered when a patient presents with any of the symptoms listed in Table 1 in the setting of loose, easily everted upper eyelids.

Treatment

The treatment of FES depends on both the severity of the patient's symptoms and, if present, the treatment of patients' underlying OSA. Conservative management may be used initially and includes eye shields and lubricating ointment at night. The shields prevent nocturnal eversion of the lids and often results in a rapid resolution of the symptoms. In addition, treatment of the sleep apnea with CPAP alone will result in resolution of the symptoms. Surgical intervention involves tightening the eyelids by removing excess tissue. Several surgical methods have been used and include full thickness wedge excision, medial/lateral tarsal strip, and medial/lateral canthal tendon plication.[5] There is evidence to suggest that the tarsal strip and canthal tendon plication surgeries show better long-term outcome in terms of symptom recurrence when compared to the full thickness wedge excision.[13] It is important to note that, because FES may be the presenting symptom in a patient with OSA, the patient should be referred for polysomnography prior to surgical intervention for FES. Surgery should be considered only once the patient's OSA is treated, otherwise the same mechanical stresses will "stretch" the patient's eyelids necessitating further treatment and surgical repair. In addition, treatment of the eyelid laxity is necessary prior to treating any coexisting eyelid abnormalities such as ptosis, as the instability of a lax lid may lead to unpredictable surgical results.

Diagnosis: Floppy lid syndrome

Epidemiology

- Typically found in overweight, middle aged men, but has been described in a broad range of patients
- Risk factors:
 - OSA
 - Obesity
 - Male gender
 - Hypertension
 - Hyperlipidemia
 - Chronic eye rubbing

Signs

- Easily everted upper eyelid with a soft, foldable tarsus
- Abnormal thickness of eyelid
- Rubbery consistency of superior eyelid
- Corneal findings:
 - Punctate epitheliopathy
 - Keratoconus
- Conjunctival findings:
 - Papillary conjunctivitis
- Periorbital:
 - Dermatochalasis, blepharochalasis, blepharoptosis, blepharitis, tear dysfunction, ectropion, lash ptosis

Symptoms

- Irritation/foreign body sensation
- Tearing
- Itching
- Redness
- Discharge

Treatment

- Eyelid lubrication and taping, lid shields
- Refer for treatment of underlying OSA prior to surgical repair
- Surgical horizontal eyelid shortening

Differential Diagnosis for Floppy Lid Syndrome

- Chronic conjunctivitis
 - Vernal conjunctivitis, giant papillary conjunctivitis, superior limbic keratoconjunctivits, toxic conjunctivitis
- Canaliculitis
- Involutional ectropion
- Nasolacrimal duct obstruction
- Sebaceous cell carcinoma

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