Floppy Eyelid Syndrome in a Child With Vernal Catarrh

Ameed N. Samaha, MD; Nadim T. Farah, MD; and Riad Maaluf, MD

INTRODUCTION

Floppy eyelid syndrome was first described as a separate clinical entity in 1981 by Culbertson and Ostler. The syndrome is characterized by a triad of loose upper eyelid that readily everts on elevating the lid; soft, rubbery tarsus that can be folded on itself; and diffuse papillary conjunctivitis of the upper tarsal conjunctiva. It has been classically described in overweight men. It may, however, occur in women, adults with hyperglicinemia, and nonobese adults. There is only one case of this syndrome reported in children. We report the youngest child with a history of chronic allergic conjunctivitis presenting with manifestations of floppy eyelid syndrome (FES).

CASE REPORT

A 7-year-old boy presented to our clinic in early 1995 with complaints of eye redness, intermittent stringy mucoid discharge, burning, and foreign body sensation in both eyes. His symptoms were worse during the early morning hours. Initial examination revealed bulbar and palpebral conjunctival injection, diffuse 2+ papillary hypertrophy of the upper tarsal conjunctiva in both eyes (Figure 1), and inferior corneal punctate staining. No Horner-Trantas dots, chemosis, or giant papillae were found.

With a 4-year positive history of seasonal (during the fall) palpebral and limbal vernal catarrh, we assumed another relapse of his allergic disease and started him on antiallergic topical and systemic treatment. This treatment included topical pheniramine maleate (Naphcon-A, Alcon Laboratories, Fort Worth, Tex), ketorolac tromethamine 0.5% (Acular, Allergan, Irvine, Calif), cromolyn sodium 2% drops and 4% ointment, and systemic (oral) loratadine 10 mg.

Despite 3 months of treatment, the patient’s signs and symptoms did not improve. Conjunctival swabs and scrapings for routine bacterial, fungal, and chlamydial cultures were negative and did not reveal any pathogenic organism. Topical steroids (prednisolone acetate and dexamethasone) were also tried to no avail.

During the course of treatment, recurrent internal and external hordeola of both upper lids were noted (Figure 2) and were conservatively managed with warm compressors and lid hygiene using fusidic acid eye ointment.

An examination 4 months after his initial presentation revealed flaccid, lax, and easily evertable upper lids (Figure 3). The upper lids could be everted without popping out, that is, without feeling the abrupt eversion of the tarsal plate signifying the loss of its rigidity. When alerted, the parents noted spontaneous lid eversion during their son’s sleep. The patient was reported to sleep alternatively on either side, but never face down.

The patient was then considered to have FES and managed by patching the lids during sleep and discontinuation of all eye drops. He responded quickly and was symptom free for the following 7 months of follow up.
DISCUSSION

The incidence of FES among patients with chronic papillary conjunctivitis was reported to be 2% in one study performed by Rapoza and colleagues. It is mainly a disease found in obese men. However, in a recent report by Culbertson and Tseng, of 60 patients with FES, only 29% were obese men and 37% were women. The pathogenesis of conjunctival inflammation is believed to be due to the eversion of the upper lid during sleep resulting in direct contact of the tarsal conjunctiva with the bedding or surface exposure. Patients who sleep on one side only characteristically have unilateral presentation.

The other explanation is the poor contact between the lax lid and the globe, also referred to as the lax lid syndrome by Van den Bosch. Patients usually present with symptoms of chronic ocular irritation, foreign body sensation, mucous discharge, and red eye. Symptoms are worse in the morning after awakening. On examination, the upper lids are flaccid, lax, rubbery, and easily evertable if pulled on without popping out. The upper tarsal conjunctiva shows diffuse papillary reaction.

Examination of conjunctival scrapings revealed keratinization of the epithelium and polymorphonuclear leukocyte infiltration. Histologic examination of the lids using light and electron microscopy and immunohistochemistry demonstrated decreased amount of elastic fibers in the tarsus as compared with the control group.

No abnormalities were noted in the collagen component of these lids. Chronic nongranulomatous inflammation in the conjunctiva and the lamina propria, as well as meibomian glands abnormalities, granuloma formation or cystic degeneration, and squamous metaplasia have also been reported.

Many speculative etiologies for the lid laxity have been suggested. These include mechanical factors, metabolic abnormalities, and degenerative processes. The coexistence of floppy eyelids and keratoconus supports the mechanical theory. Moreover, in some reports, symptoms and signs decreased when a patient’s body weight decreased.

The associations of FES with hyperglyceminia and with the sleep apnea syndrome may suggest a connective tissue disorder as a possible etiologic factor. The similarities between lax lids and blepharochalasia led some to believe that these two entities, in fact, represent two manifestations of the same degenerative process.

In our patient, excessive rubbing of the eyelids caused by the severe itchy sensation during the active phases of vernal catarrh could be incriminated as a possible mechanical factor leading to the lid laxity and flaccidity. This is supported by the histological findings of Netland and colleagues who found that eyelid rubbing and mechanical pressure may mechanically or enzymatically degrade the eyelid elastic fibers leading to lax lids.

Overweight and face-down sleeping were not demonstrated as risk factors in our patient. The presentation was that of chronic papillary conjunctivitis refractory to medical treatment. The diagnosis was established late because of the previous history of vernal catarrh, and palpebral opacity was not checked before.

The inferior punctate corneal staining is partly attributed, in addition to exposure, to the qualitative tear film disturbances caused by meibomian glands dysfunction and abnormalities associated with FES, which manifested as recurrent hordeola in our case. Knowing that lid taping per se is not expected to improve the quality of the tear film or the function of the meibomian glands, we attribute most of the patients symptoms to the laxity of his lids.

Management of FES includes conservative or surgical treatment. Conservative treatment consists of patching or taping the lids or the use of eye shields during sleep. Effectiveness of such treatment is evident in the first few weeks.

As in papillary conjunctivitis, specifically vernal catarrh, we would initially rely on the subjective response of the patient (improvement of symptoms and eye redness), as some of the ocular signs (papillae) require months to resolve.

If conservative treatment fails, horizontal shortening or thickening of the upper lid through different techniques are the mainstay of surgical treatment. Our patient stayed symptom free for 7 months after patching his eyelids during sleep. His condition was not severe enough to warrant surgical intervention.

FES may complicate the course of allergic conjunctival diseases of the eyes, especially those with
more severe and prolonged manifestations, such as vernal catarrh. We believe that excessive eye rubbing is an important mechanical factor leading to lid laxity in such patients. These patients usually present with chronic papillary conjunctivitis refractory to medical treatment. Simple external examination is sufficient for the diagnosis of this entity. FES should be considered in the differential diagnosis of chronic conjunctivitis in children.

REFERENCES