The lax eyelid syndrome

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Abstract

The floppy eyelid syndrome (FES) was first described in middle aged, obese men. In later descriptions, age and sex were not specifically mentioned. Associations of FES with various other syndromes have been described. The authors question whether all these cases represent the same, single, syndrome. They suggest that a clinical picture similar to FES may occur in lax upper eyelids of any cause. Four such cases are reported here. The authors therefore coin the more general term 'lax eyelid syndrome'. They suggest using the term 'floppy eyelid syndrome' uniquely for patients with the classic signs and symptoms.

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Culbertson and Ostler first described the floppy eyelid syndrome (FES) in 1981.¹ The syndrome was characterised by a 'floppy' – that is, easily evertible, upper eyelid, and a papillary conjunctivitis of the upper palpebral conjunctiva. Patients often presented with a long history of irritation and discharge. The syndrome only occurred in middle aged, obese men. Unnoticed eversion of the upper eyelid during sleep was thought to cause the papillary conjunctivitis. At first, effective treatment consisted of shielding the affected eye at night.¹ Later, surgical correction of upper and, in some cases, lower eyelid laxity by means of full thickness eyelid shortening proved to be very effective.²⁻⁴ Interestingly, later papers described similar signs and symptoms in young or non-obese patients, or in association with various disorders such as keratoconus, sleep apnoea syndrome, Meibomian gland dysfunction, and the blepharochalasis syndrome.⁵⁻¹⁰ Despite these departures from the

original description, the affection of all patients was none the less diagnosed as FES. Its cause, however, remained obscure.

In this paper, four patients are described who had several signs and symptoms similar to those described in FES. All of them had an upper eyelid laxity with varied, albeit clearcut, causes. We shall question whether the term FES applies, or whether a more general term should be adopted.

Case reports

CASE 1

An 86-year-old woman (height 160 cm, weight 51 kg) had presented elsewhere with a history of chronic discharge and irritation of the left eye for more than a year. The symptoms showed no diurnal variation. She did not report sleeping with the affected side on the pillow, nor had she ever noticed upper eyelid eversion during sleep. On examination, her left eye had shown purulent discharge, a papillary conjunctivitis, and a diffuse punctate keratitis. She had contracted a moderate ectropion of the left lower eyelid. No corneal vascularisation had been present. A culture from the conjunctiva had grown *Staphylococcus aureus*. She had been treated with several topical antibiotics. Because of a high intraocular pressure, timolol eyedrops also had been prescribed. Despite several weeks of continued treatment, her symptoms had not improved. She was then referred to our oculoplastic service, where we corrected her lower eyelid ectropion by excising a 7 mm full thickness block from the lateral part of her eyelid margin. Thereafter, her symptoms diminished markedly. After 2 years, however, the discharge

Fig 1A

Figure 1 (A) Case 1 at first presentation. The affected left eye is closed. (B) The upper eyelid is pulled laterally. Conjunctival changes and severe discharge are visible. The nasal sclera is not visible, owing to laxity of the medial canthal tendon.

Fig 1B

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Fig 2A

Fig 2B

Figure 2 (A) Case 1, 1 month after excision of a full thickness block from the left upper eyelid and shortening the posterior lamella of the left lower eyelid under a skin muscle flap. (B) When the upper eyelid is lifted, no conjunctival injection is visible.

and keratitis of the left eye recurred. This time, both the medial canthal tendon and the lateral canthal ligament were very lax. Surgical reinsertion of both was performed. This resulted clinically in a better apposition of both upper and lower eyelid against the globe. Within a few weeks after the operation her signs and symptoms improved dramatically. Two years later, at age 90, she presented for the third time with severe discharge and a diffuse punctate keratitis (Fig 1).

On examination, she showed a recurrence of the laxity of her medial canthal tendon and lateral canthal ligament. We treated her upper eyelid laxity by removing an 11 mm full thickness block from the temporal part of the lid. To treat her lower eyelid laxity, we excised a block of posterior lamella in the lateral canthal angle under a skin muscle flap. In the following weeks, the discomfort, keratitis, and discharge disappeared. A slight left upper eyelid ptosis remained. She has been free of the symptoms for 6 months (Fig 2).

Histology of the excised upper eyelid block showed severe, active, and chronic conjunctivitis with bacterial colonies on the conjunctival surface. The tarsal plate showed moderate to severe lipomatous atrophy. In the Meibomian glands, the mite *Demodex brevis* was present.

CASE 2

An 84-year-old man (height 170 cm, weight 56 kg) was referred to our oculoplastic service because of a punctate epithelial keratitis of his right eye, attributed to an upper eyelid entropion. He reported that he had been having discharge from the eye, and a gritty feeling, for more than 6 months. Symptoms were worse in the evening. He did not report eyelid eversion during the night. Elsewhere, his symptoms had been ascribed to a coexisting blepharitis. He had

been treated with eyelid massage, povidone iodine eyedrops, and eyedrops containing trimethoprim and polymyxin B for several months, without any improvement. When we examined him, the lashes of the right upper eyelid showed a ptosis. However, they did not touch the cornea. In addition, his right upper and lower eyelid proved to be very lax (Fig 3).

There was no excessive laxity of the medial canthal tendon or of the lateral canthal ligament. The eyelid laxity appeared to be due to the combined effects of involutional changes of the tarsal plate and to an age-related enophthalmos caused by atrophy of orbital fat. On eversion of the upper eyelid, the tarsal plate had a normal consistency. A moderate papillary conjunctivitis of the tarsal conjunctiva was present. He also had an ulcerative blepharitis. There was a slight injection of the bulbar conjunctiva, together with a diffuse punctate keratitis. The upper eyelid laxity was corrected by excision of a 15 mm full thickness block of the temporal part of the lid and the lower eyelid laxity was treated by excision of a 5 mm full thickness block in the lateral canthal angle. Two weeks after surgery, the patient was free of the symptoms. The punctate keratitis and the blepharitis disappeared fully within a month. Interestingly, after correction of the horizontal laxity, the lash ptosis also disappeared. He has been free of symptoms now for 18 months (Fig 4).

Histological sections of the excised upper eyelid block showed a moderate subconjunctival, chronic, inflammatory infiltration. In the tarsal plate a lipogranulomatous inflammation was present. *Demodex brevis* was demonstrated in the

Fig 3A

Fig 3B

Figure 3 (A) Case 2 before surgery. (B) When the eyelid is pulled laterally, the severe laxity of the right upper eyelid is visible.

Figure 4 Case 2, 2 months after excision of a full thickness block from right upper and lower eyelid.

Meibomian glands. The dermis showed severe elastic degeneration.

CASE 3

An 83-year-old, moderately obese woman (height 165 cm, weight 82 kg) was referred to our oculoplastic service because of punctate epithelial keratitis of the right eye, thought to be due to a facial palsy. The facial palsy had been present since an operation of the mastoid bone at the age of 38. She had never undergone any surgery to improve eyelid apposition or function. For several weeks she had suffered from strong irritation, discharge, and tearing from her right eye. Her complaints did not vary during the day. She had not experienced any nocturnal eyelid eversion. Treatment with lubricant eyedrops had not been satisfactory. When we examined her, the right lower eyelid showed an ectropion with keratinisation of the eyelid margin. Her right upper eyelid was slightly ptotic. On forceful squeezing, her upper eyelid fully covered the cornea, although the eyelids did not close completely, because of the lower lid ectropion. The conjunctiva of the affected eye was injected, and the cornea showed punctate epithelial keratitis of the lower and central part, as in exposure keratitis due to facial palsy. We repositioned her lower eyelid through a lateral tarsal strip procedure. Postoperatively, a slight ectropion of the eyelid margin persisted, but eyelid closure was adequate. However, her signs and symptoms did not improve (Fig 5). On further investigation, the upper eyelid appeared to be moderately lax, and the tarsal plate of the upper lid showed a papillary conjunctivitis.

The upper eyelid laxity was then treated by excision of a 7 mm full thickness block. Unlike typical cases, the wound touched the cornea postoperatively. Nevertheless, the symptoms disappeared within 2 weeks. The ptosis also disappeared. Although the lower lid ectropion has recurred slightly, she has been free of symptoms for 8 months (Fig 6).

Fig 5A

Figure 5 (A) Case 3, 3 weeks after correction of right lower lid ectropion, before correction of upper lid laxity. (B) Moderate laxity of the right upper eyelid is visible when the eyelid is pulled laterally.

Histological slides of the excised upper eyelid block showed a subconjunctival chronic follicular inflammation. In the tarsal plate, a severe lipomatous atrophy of the Meibomian glands was noted. There was a dense, chronic inflammation around the accessory lacrimal tissue.

CASE 4

A 66-year-old, frail woman (height 156 cm, weight 39 kg) had undergone a cataract extraction of the left eye elsewhere 18 months before she was referred to our hospital. A few days after the cataract extraction, she had developed a corneal ulcer. About the same time, a ptosis of the left upper eyelid had come about. Treatment with topical antibiotics had cured the corneal ulcer, although stromal loss of the lower part of the cornea had remained. Since the cataract extraction, she had been suffering almost permanently from a purulent conjunctivitis, for which she had been treated unsuccessfully with various topical antibiotics. Finally, she was referred for evaluation of the chronic conjunctivitis. She complained of chronic discharge and irritation without diurnal variation. She had not experienced eyelid eversion during sleep. When we examined her, her left upper eyelid showed a ptosis with a vertical eyelid fissure of 3.5 mm. The levator function was 12 mm and the upper lid skin crease position was high, suggesting that the ptosis was due to a disinsertion or thinning of the levator aponeurosis (Fig 7).

Both upper and lower eyelids were very lax. The temporal sclera was almost fully covered by the eyelids, probably because of laxity of the lateral canthal ligament. The lower eyelid showed a medial ectropion. On eversion of the upper eyelid, the consistency of the tarsal plate was normal. The tarsal conjunctiva showed a slight papillary conjunctivitis. A blepharitis coexisted on her upper and lower eyelids. Inspection of the cornea revealed a diffuse punctate epithelial keratitis, in addition to the stromal loss and vascular ingrowth of the lower part. The bulbar conjunctiva was moderately injected.

Her blepharitis was treated successfully with eyelid scrubbing and application of 1% fusidic acid eye gel on the eyelids. Nevertheless, the keratitis remained during the following 3 months. We then corrected the laxity of upper and lower eyelid by reinserting the lateral canthal ligament to the orbital rim. At the same time we treated her ptosis through a monitored anterior levator reinsertion. On postoperative examina-

Fig 5B

Figure 6 Case 3, 4 months after excision of a full thickness block of the right upper eyelid. Although the lower lid ectropion has recurred slightly, no conjunctival injection is visible.

tion, the reinsertion of the lateral canthal ligament had caused a much tighter apposition of both upper and lower eyelid against the globe. Although some overcorrection of the upper eyelid ptosis existed, the patient was pleased with the result. After the operation, the punctate keratitis and conjunctival redness disappeared within 2 weeks. She has been free of symptoms without the use of any eyedrops for 6 months.

Discussion

To date, FES has not been clearly defined in the literature. Culbertson and Ostler described it as 'an idiopathic disease occurring in middle-aged, obese men, characterised by an easily everted, floppy upper eyelid and upper palpebral papillary conjunctivitis'.¹ However, later definitions did not include the original criteria of age, sex, and the altered consistency of the tarsal plate.⁵⁻⁸ Whatever its exact definition, the cause of the syndrome has remained unclear. Nevertheless, associations between symptoms of FES and various other clinical syndromes have been described, such as the sleep apnoea syndrome, keratoconus, Meibomian gland dysfunction, chronic conjunctivitis, and the blepharochalasis syndrome.⁵⁻¹⁰ Again, all these reports provide different descriptions of FES. One could therefore question whether all these cases of FES represent the same clinical entity. Therefore, any speculations on the possible causes of FES should be viewed with caution.

Contrary to most of the literature on FES, there was a clearcut cause for the eyelid laxity in each of our four presented cases. In our first case, the eyelid laxity was caused by involution of the medial canthal tendon and the lateral canthal ligament. Recurrence of the symptoms was

related to recurrence of eyelid laxity. In the second case, involutional changes of the tarsal plate and atrophy of orbital fat accounted for the upper eyelid laxity. In the third case, both the upper eyelid laxity and lower eyelid ectropion could be attributed to the combined effects of involution and a paretic orbicularis muscle. While lower eyelid repositioning failed to cure this patient, upper eyelid shortening did. This suggests a causative role of upper eyelid laxity. In the last case, both the ptosis and the disinsertion of the lateral canthal ligament were probably caused by either traction on the eyelids during cataract extraction, or by excessive squeezing as a result of the photophobia induced by a corneal ulcer.

Of interest is the alleged possible role of tear film disorders in the pathogenesis of FES. One paper reported on seven patients with both FES and chronic conjunctivitis. Three of these patients showed evidence of both quantitative and qualitative tear film disorders.⁵ In another series, all three patients with FES had an inadequate tear film.⁹ Another paper reported on the histology of the tarsal plate of a patient with FES. Cystic degeneration and squamous metaplasia of the Meibomian glands were found, probably causing qualitative tear film abnormalities.¹⁰

In each of our four cases, a coexisting tear film disorder may have been present. Microscopy of the excised upper eyelid blocks revealed that the Meibomian glands of patients 1 and 2 were infested with *Demodex brevis*. The tarsal plate of patients with the symptoms of FES is frequently infested by *Demodex brevis*.¹² This mite may cause a tear film disorder through destruction of the Meibomian glands.^{10,11} On microscopy, the upper eyelid tarsal plate of case 3 showed a strong lipomatous atrophy of the Meibomian glands. On clinical examination, cases 2 and 4 showed a blepharitis which might have been associated with Meibomian gland loss.¹³ In conclusion, all four patients probably had tear film disorders. However, they were cured by improving the apposition force between the eyelids and the globe. This was achieved by upper and, if necessary, lower eyelid shortening in the first three cases, and by reinserting the lateral canthal ligament in the last case. Similar results have been reported in the literature.^{5,10} Since eyelid shortening cannot be expected to improve tear film quality, tear film disorders can, at most, only be regarded as contributing factors in the pathogenesis of FES.

Fig 7A

Fig 7B

Figure 7 (A) Case 4 at presentation. (B) Two months after ptosis correction and reinsertion of the lateral canthal ligament.

Taken together, we would argue that eyelid laxity of any cause is the most important causative factor of a clinical picture resembling FES. This picture consists of a papillary conjunctivitis, punctate epithelial keratitis, and ocular discharge. We would speculate that a drop in the apposition force between the eyelids and eyeball below a critical level probably might cause these signs and symptoms. Furthermore, any coexisting tear film abnormalities might luxate or aggravate the clinical picture. Such tear film abnormalities could not singularly account for the entire clinical picture, because that would not explain the success of upper eyelid shortening. In our experience, many elderly patients have a similar eyelid laxity, without any symptoms. Therefore, other not yet identified factors may play a role in the aetiology of this clinical picture. We contend that the middle aged, obese men with a rubbery tarsal plate originally described by Culbertson and Ostler comprise a typical and easily identifiable subgroup within the group of patients with signs and symptoms related to upper eyelid laxity. Unfortunately, such symptoms resulting from upper eyelid laxity are often thought to be limited to this subgroup. As a result, many patients will presumably not be diagnosed and treated correctly. Therefore, we suggest reserving the term 'floppy eyelid syndrome' (FES) to the originally described subgroup. For patients who have a chronic papillary conjunctivitis, punctate epithelial keratitis,

and ocular discharge related to upper eyelid laxity the term 'lax eyelid syndrome' (LES) should be introduced. We hope that this distinction will lead to an earlier recognition, and subsequently, more adequate and timely treatment of patients suffering from lax eyelids of any cause.

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