Management of Bilateral Congenital Upper Eyelid Eversion with Severe Chemosis

Oluyemi Fasina, FWACS

Department of Ophthalmology, University College Hospital / University of Ibadan, Ibadan, Oyo State, Nigeria

Purpose: To report a case of complete bilateral congenital upper eyelid eversion associated with severe chemosis in a newborn, and to describe a semi-invasive technique for its management.

Case Report: The patient was a four-hour-old Nigerian neonate with bilateral congenital upper eyelid eversion with severe and progressive chemosis. Conservative management failed to resolve the condition. However, compression eyelid sutures resulted in prompt and satisfactory resolution.

Conclusion: Compression eyelid suturing is a semi-invasive technique for management of severe chemosis due to congenital upper lid eversion resulting in rapid and satisfactory resolution of the condition.

Keywords: Congenital; Eyelid Eversion; Lid Suture; Management

INTRODUCTION

Congenital upper eyelid eversion has been defined as protrusion of the palpebral conjunctiva from an everted eyelid.1 This rare condition,1,5 was first described and denominated as “double congenital ectropion” in 1896 by Adams6. It typically presents bilaterally, and can sometimes be acquired following surgical correction of congenital ptosis.7 The eversion is usually present at birth, but late-onset cases have also been reported.8 The incidence of congenital upper eyelid eversion appears to be higher in black infants, cases with trisomy 21 and neonates with collodion skin disease.8-10 Due to its rarity, the incidence of the condition is unknown.11 Treatment is largely conservative, and resolution is expected over days to weeks. Surgical management, however, is indicated in some cases.

Herein, we report a case of complete bilateral eversion of the upper eyelids in an otherwise normal black infant, associated with severe and progressive prolapse of chemotic palpebral conjunctiva. While the patient was primarily unresponsive to conservative management, compression eyelid sutures provided immediate and satisfying resolution of the condition.

CASE REPORT

A Nigerian male neonate was referred from a private hospital four hours after uncomplicated spontaneous vaginal delivery with bilateral fleshy masses protruding out of both eyes together with eversion of both upper eyelids. The condition had been noticed immediately after birth and gradually increased after the child started crying (Fig. 1). He was the second child of a 30-year-old mother and product of a full-term uneventful pregnancy. There was no
history of genital infections during the perinatal course, neither instrumentation at delivery.

After instillation of a topical anesthetic agent, insertion of Desmarres retractors and retraction of the everted upper eyelids, examination revealed grossly normal, non-proptotic eyeballs with clear corneas, anterior chambers of normal depth, and round, central and reactive pupils. No other abnormality was noted.

Conservative management was tried by manually repositioning the eyelids and applying pressure patches which was unsuccessful due to severe chemosis. Because of the high possibility of traumatizing the severely chemotic conjunctiva, tarsorrhaphy was considered to be technically difficult in this case; therefore, compression eyelid sutures were applied as described below.

Twenty minutes after topical application of an anesthetic ointment (lidocaine/prilocaine; Prilox cream, Neon Laboratories, Mumbai, India) to the eyelids and cheeks, 4-0 polypropylene sutures were passed through the upper eyelid margins, the chemotic conjunctiva was tucked in using blunt forceps, and both upper eyelids were sutured to the corresponding cheeks (Fig. 2). Antibiotic ointment was instilled and a light dressing was applied over the lids.

After 36 hours, conjunctival protrusion was reduced considerably. Subsequently, the sutures were removed and the eyelids remained reverted with minimal conjunctival chemosis (Fig. 3). The patient was placed on twice-daily antibiotic ointment and instructed to return one week later. The parents were instructed to return if eyelid eversion recurred any time.

After one week, there was complete resolution of chemosis and the lids remained reverted (Fig. 4). The child could open his eyes normally and all other ocular examinations were within normal limits. After taking a detailed

Figure 1. Appearance of the four-hour-old neonate at presentation; complete bilateral eversion of the upper eyelids is present together with severe conjunctival chemosis and prolapse.

Figure 2. “Compression” eyelids sutures in place.

Figure 3. Lid sutures were removed after 36 hours; note the markedly reduced chemosis and reverted eyelids.

Figure 4. The same baby one-week after intervention; the eyelids are well-reverted.
review by a pediatrician no other abnormality was detected. The lids remained reverted at five weeks and the conjunctiva was non-chemotic and normally positioned (Fig. 5).

DISCUSSION

Complete eversion of the upper eyelids with conjunctival chemosis and prolapse is a rare condition in neonates. The underlying cause of the disease remains largely unknown, yet several possible mechanisms have been postulated. Birth trauma, orbicularis muscle hypotonia, vertical shortening of the anterior eyelid lamella or vertical elongation of the posterior eyelid lamella, lateral elongation of the lid, failure of the orbital septum to fuse with the levator aponeurosis, and absence of an effective lateral canthal ligament have been implied as probable underlying causes.\(^2\),\(^12\),\(^13\) It has been suggested that pressure exerted on the baby as it passes through the birth canal, may induce venous stasis, leading to conjunctival chemosis and prolapse, thereby causing eversion of the eyelid.\(^13\) Additionally, the resulting orbicularis spasm may function as a sphincter in the everted eyelid, leading to a vicious cycle of conjunctival strangulation and edema, further worsening the condition.\(^13\) Histopathological examination of the everted eyelids in an infant who died 9 days after birth by Young did not reveal any abnormality.\(^14\) Regardless of the etiology, the eversion-chemosis cycle should be interrupted. The chemotic conjunctiva usually protects the cornea from exposure and so far, only one case of corneal perforation due to congenital eversion of the eyelids has been reported.\(^2\)

Treatment is aimed at protecting the exposed conjunctiva from desiccation and infection. Mild to moderate cases may resolve spontaneously by applying moist dressings, eyelid taping, and pressure patching along with topical antibiotic and anti-inflammatory lubricants. Additionally, topical hypertonic saline may be employed as a conservative treatment.\(^4\),\(^15\) These treatment modalities however, take days or weeks to resolve the chemosis and eversion.

Certain factors led us to employ the semi-invasive treatment modality described herein. Socio-cultural values attached to the condition in our environment and the psychological impacts on the parents, often hinder conservative management.\(^4\) The parents occasionally apply unsafe practices especially when the neonates are managed on an out-patient basis leading to potentially undesirable consequences.\(^16\) Conservative management of such babies may require prolonged hospital admission, frequent hospital visits and negative economic effects. Other treatment modalities like subconjunctival hyaluronic acid injection were not available in our setting and were not likely to be effective with the severe chemosis in our patient.\(^17\)

Surgical treatment is usually indicated for non-resolving cases or those presenting late. These include temporary tarsorrhaphy, fornix sutures, full thickness skin grafts, horizontal eyelid shortening and excision of redundant conjunctiva.\(^1\)

Due to the presence of severe chemosis and failure of eyelid reversion with patching, we managed our patient with compression eyelid sutures. Mechanical compression of the chemotic conjunctiva and “forcing out” fluid was the probable mechanism through which the lid sutures acted. It also helped in stretching the lid, thereby elongating the anterior lid lamella. Reluctance of the parents to wait for a few days or weeks for spontaneous resolution of the condition through conservative management, and social stigma attached to the condition necessitated this surgical approach in our case.

In summary, congenital eyelid eversion is a rare condition which is generally treated conservatively. Nevertheless, socio-cultural and
economic factors may not allow the relatively long period required for gradual resolution of the condition. Any treatment modality that resolves the severe and progressive chemosis within a day or two will be preferred by both physicians and parents. Besides, the risk of desiccation and infection would be markedly reduced and hospital admission would not be necessary. Compression eyelid sutures appear safe and result in faster resolution of conjunctival chemosis with satisfying outcomes. Hence, this treatment approach is suggested in cases with complete congenital eversion of the upper eyelids associated with severe chemosis.

Conflicts of Interest

None.

REFERENCES

1. Shinder R, Langer PD. Unilateral congenital eyelid eversion causing marked chemosis in a newborn. J Pediatr Ophthalmol Strabismus 2010;1:1-2.
2. Al-Hussain H, Al-Rajhi AA, Al-Qahtani S, Meyer D. Congenital upper eyelid eversion complicated by corneal perforation. Br J Ophthalmol 2005;89:771.
3. Maheshwari R, Maheshwari S. Congenital eversion of upper eyelids: case report and management. Indian J Ophthalmol 2006;54:203-204.
4. Adeoti CO, Ashaye AO, Isawum MA, Raji RA. Non-surgical management of congenital eversion of the eyelids. J Ophthalmic Vis Res 2010;5:188-192.
5. Cuvuoto KM, Hui JL. Congenital eyelid eversion. J Pediatr Ophthalmol Strabismus 2010;47 Online: e1-3.
6. Adams AL. A case of double congenital ectropion. Med Fortnightly 1896;9:137-138.
7. Wolfley DE. Preventing conjunctival prolapse and tarsal eversion following large excisions of levator muscle and aponeurosis for correction of congenital ptosis. Ophthalmic Surg 1987;18:491-494.
8. Sellar PW, Bryars JH, Archer DB. Late presentation of congenital ectropion of the eyelids in a child with Down syndrome: a case report and review of the literature. J Pediatr Ophthalmol Strabismus 1992;29:64-67.
9. Lu LW, Bansal RK, Katzman B. Primary congenital eversion of the upper lids. J Pediatr Ophthalmol Strabismus 1979;16:149-151.
10. Shapiro RD, Soentgen ML. Collodion skin disease and everted eyelids. Postgrad Med 1969;45:216-219.
11. Duke Elder S. Duke Elder’s System of Ophthalmology. St. Louis: C.V. Mosby; 1964.
12. Blechman B, Isenberg S. An anatomical etiology of congenital eyelid eversion. Ophthalmic Surg 1984;15:111-113.
13. Raab EL, Saphir RL. Congenital eyelid eversion with orbicularis spasm. J Pediatr Ophthalmol Strabismus 1985;22:125-128.
14. Young RJ. Congenital ectropion of the upper lids. Arch Dis Child 1954;29:97-100.
15. Isawumi MA, Adeoti CO, Umar IO, Oluwatimilehin IO, Raji RA. Congenital bilateral eversion of the eyelids. J Pediatr Ophthalmol Strabismus 2008;45:371-373.
16. Fasina O, Ubah JN. Pattern of pre-hospital consultation among ophthalmic patients seen in a tertiary hospital in South West Nigeria. Afr J Med Med Sci 2009;38:173-177.
17. Taban M, Mancini R, Nakra T, Velez FG, Ela-Dalman N, Tsirbas A, et al. Nonsurgical management of congenital eyelid malpositions using hyaluronic acid gel. Ophthal Plast Reconstr Surg 2009;25:259-263.