Congenital combined eyelid imbrication and floppy eyelid syndrome: Case report and review of literature

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Congenital eyelid imbrication syndrome (CEIS) is an extremely rare, benign, transient, self-limiting eyelid malposition disorder. The classic triad of signs in patients with a CEIS consists of bilateral upper eyelids overriding the lower eyelids when child was in sleep, bilateral medial and lateral canthal tendon laxity and tarsal conjunctival hyperemia. We report a third case of congenital combined eyelid imbrication and floppy eyelid syndrome in healthy neonate that was resolved within a week with conservative treatment.

Key words: Congenital ectropion, congenital eyelid imbrication syndrome, congenital floppy eyelid syndrome, congenital lax upper eyelid syndrome, down syndrome

Eyelid imbrication syndrome is an idiopathic eyelid malposition disorder and characterized by upper eyelids overriding the lower eyelids.[1] In adults, eyelid imbrication is usually

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Manuscript received: 12.12.12; Revision accepted: 12.07.13

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/0301-4738.121086

Video files available on www.ijo.in

associated with floppy eyelid syndrome and required surgical correction. Congenital eyelid imbrication syndrome (CEIS) is extremely rare, benign and self-liming condition and sometime it may associate with congenital floppy eyelid syndrome (CFES).[2] According to a computerized MedLine and Google search, only three cases of CEIS, out of them two were associated with CFES has been described in the literature.[2-4] We report a case of CEIS associated with CFES that resolved with conservative treatment.

Case Report

A full-term newborn Indian female, after caesarean section, was referred to us 16 h after delivery for eye discharge, inability to open eyelids completely and abnormal overriding of upper eyelids on lower eyelids that was observed immediately after birth. The parameters at birth were as follows: Birth weight 3200 g, length 48 cm, head circumference 38 cm and Apgar score 10/10. She was the first child in the family. There was no family history of ocular malformation, syndromes or consanguinity. Retrospective review of antenatal record did not revealed intake of any teratogenic drug.

Ocular examination of child asleep showed elongated, bulky upper eyelids and upper eyelids overlapping on lower eyelids, 6 mm OU [Video 1], [Fig. 1]. The child having moderate mechanical, blepharoptosis on both sides in the awakened state. The ocular anthropometric measurements on the first postnatal day were as following: Vertical palpebral fissure in awakening state 5 mm, horizontal palpebral length 18 mm, horizontal length of upper eyelid 24 mm, and midpoint vertical upper eyelid height 10 mm in both eyes. The canthal tendons of both eyelids are symmetrically laxed and round in appearance. The upper eyelids were easily everted by gentle pulling of the skin of the upper eyelid toward the forehead [Fig. 2]. The both upper eyelids had a tendency for spontaneous complete eversion on crying and yawing [Fig. 3]. The lids required manual repositioning to the normal position. Tarsal conjunctiva of both upper eyelids shows mild hyperemia. Both cornea were clear and did not show staining with rose-bengal or fluorescein stain. Upper eyelid margins were faintly stained with rose-bengal staining. Tactile intraocular pressure seems to be within the normal range. Both eyes pupillary reactions are brisk. The rest of adnexal, anterior segment and dilated fundus examination were unremarkable. Discharge from eyelids and cul-de-sac was sent for culture and sensitivity. No features of Down syndrome or sleep apnea syndrome were found on systemic evaluation. The newborn was treated with topical gatifloxacin 0.3%, QID; carboxymethyl cellulose 1%, 6 times a day and 1% tetracycline eye ointment, QID. She was followed on a daily basis. The next day culture of conjunctival and eyelid discharge grew *Staphylococcus aureus*, sensitive to ampicillin, ciprofloxacin, gatifloxacin, tobramycin and gentamycin. The newborn was switched to tobramycin eye drop and ointment as well. B-scan eyeball and orbit showed normal findings. Correction 4 mm overriding of eyelids was noted on both sides within first 48-h of the postnatal period. At 7th postnatal day child's eyelids position became normal without any residual blepharoptosis [Fig. 4].

Discussion

CEIS is a rare eyelid malposition disorder and characterized by overriding of upper eyelid eye on lower.[1] Acquired imbrication eyelid syndrome is common in adults above the age of 40 years and usually seen in patients who underwent lateral tarsal strip procedure for lower eyelid laxity and floppy eyelid syndrome.[1] CEIS is frequently associated with CFES.[3,4] It is characterized by overriding of upper eyelids on lower eyelids, apparent congenital entropion of both lower eyelids and tarsal conjunctival hyperemia. CEIS is a self-limiting disorder and did not require any surgical intervention.[2,3] Instillation of artificial tear substitutes may be helpful in relieving the foreign body sensation from skin and eyelashes. Floppy eyelid syndrome is frequently associated with Down syndrome.[5] In the present case, no features of Down syndrome were noted on systemic evaluation. Our case is different from previously reported three cases because of associated ophthalmia neonatorum and blepharoptosis [Table 1]. In the present case, overriding of upper eyelid on the lower eyelid is also the most severe than previously reported cases.[2-4] Our child also having moderate mechanical blepharoptosis, which was disappeared within 1 week. Spontaneous tightening of

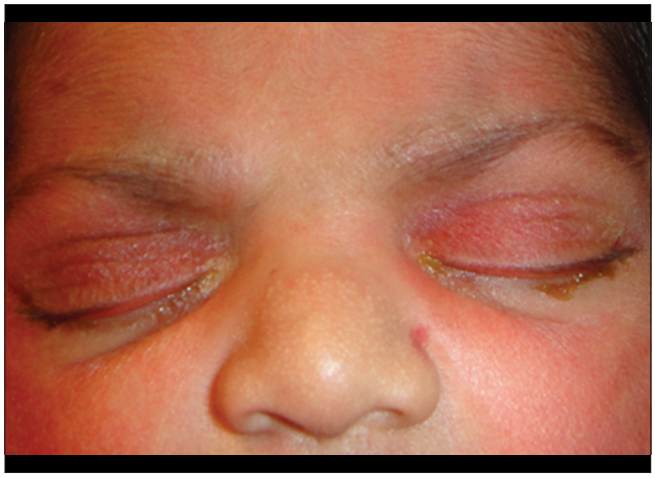

Figure 1: Clinical photograph of neonate with bilateral congenital eyelid imbrication and floppy eyelid syndrome showing overlapping of upper eyelids on lower eyelids on eye closure. The eyelashes of the both lower eyelids are completely covered by the upper eyelids.

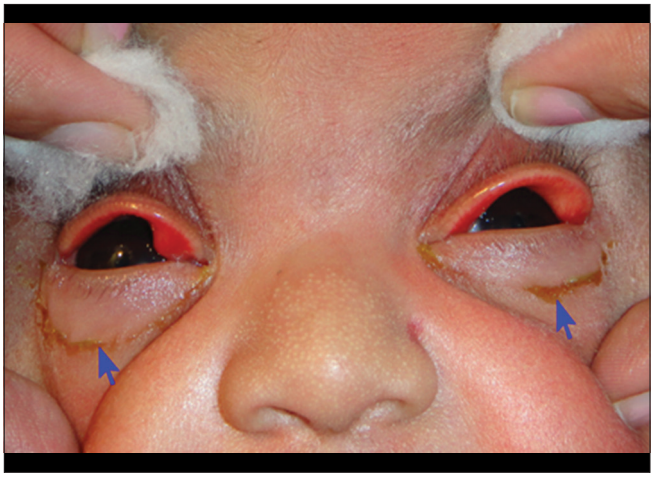

Figure 2: Clinical photograph of neonate with bilateral congenital eyelid imbrication and floppy eyelid syndrome shows eversion of upper eyelids with minimum efforts. Blue arrows indicate discharge on the lower eyelid and marking the extent of overriding.

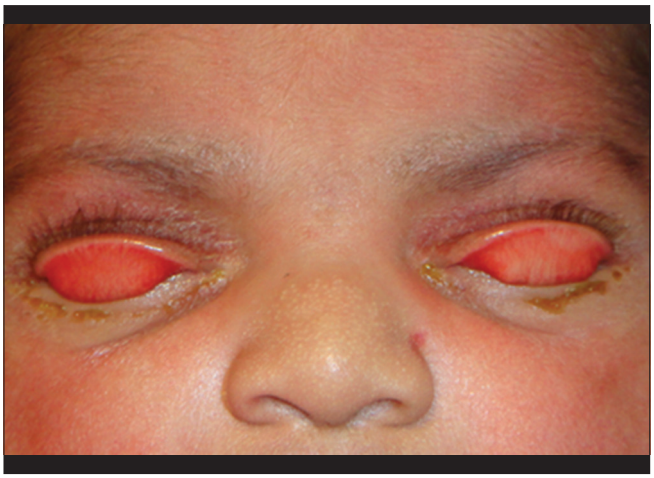

Figure 3: Clinical photograph of neonate with bilateral congenital eyelid imbrication and floppy eyelid syndrome showing spontaneous eversion of both upper eyelids while yawing and tarsal conjunctival mild hyperemia.

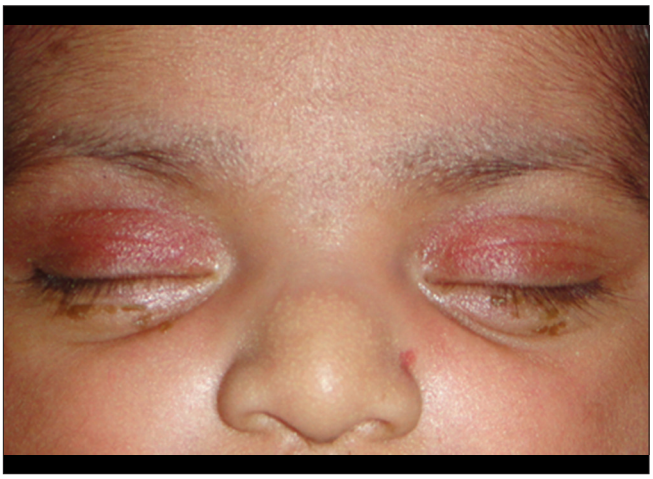

Figure 4: Clinical photograph of neonate with bilateral congenital eyelid imbrication and floppy eyelid syndrome showing complete resolution of overlapping of upper eyelids on lower eyelids and laxity of upper eyelid medial and lateral canthal tendon.

Table 1: Clinical presentation of reported cases of congenital eyelid imbrication syndrome

Features	Rumelts *et al.*[2]	De Silva *et al.*[3]	Odat *et al.*[4]	Present case
Age at presentation	3 h	48 h	>24 h	16 h
Sex	Female	Male	Male	Female
Method of delivery	Vaginal	NM	Vaginal	Caesarean section
Amount of overriding	OU, 1.25 mm	OU,>1 mm	OD>OS; OS, 1-2 mm	OU 6 mm
Canthi	Laxed and longer	Laxed	Laxed	Laxed and longer
Spontaneous eversion of upper eyelids	Absent	Present	Present	Present
Purulent discharge	NM	NM	Present	Present
Papillary reaction	Absent	Few papillae	NM	Absent
Recovery	1 week	2 months	3 weeks	1 week

NM: Not mentioned, OU: Ocular Utrique, OD: Oculus Dexter, OS: Oculus Sinister

canthal tendons and involutional changes in upper eyelids during early postnatal period thought to be responsible for correction of blepharoptosis. In acquired floppy eyelid syndrome, blepharoptosis and lash ptosis is quite common and required surgical intervention.

Previous researcher thought that postnatal growth of the bony orbit may contribute to the spontaneous tightening of canthal tendons resulting in correction of the CEIS.[2] We feel postnatal growth of the bony orbit have no contribution in spontaneous correction of eyelid imbrication because orbital growth in 1 week is not so much significant to correct 6 mm of overriding of eyelids as seen in our case.[6] Moreover, 4 mm overriding correction was observed within first 48-h of the postnatal period. We observed that upper eyelids looks, elongated, floppy and odematous at day one which became erythematous and firm on day 7. We feel whole the eyelids were bulky and floppy and underwent involutional changes under the influence of unknown effect in 1st week of life and resulted in tightening of laxed canthal tendons and normalization of size and tone of upper eyelids. Spontaneous eversion of the eyelid is a classical feature of floppy eyelid syndrome. In CEIS, spontaneous eversion is directly related to the amount of overriding of upper eyelid on lower. This relationship very well-documented in Odat *et al*. and our case.[4] We feel CEIS is a less severe form of CFES rather than different entity. It may be more appropriate to use the term congenital lax upper eyelid syndrome instead of CEIS/CFES.

Conclusion

CEIS is a rare, self-limiting, transient disorder of uncertain cause. The combination of CEIS and CFES should be considered in the differential diagnosis of congenital eyelid malposition. An ophthalmologist should be familiar with CEIS and its possible complications such as corneal epithelial defect and even ulcers. The floppy eyelid syndrome should be searched in all newborns with a CEIS. This case report may expand current recognition and understanding of this rare entity.

Acknowledgment

We would like to thank the parents of the child for their cooperation and for consenting to the publication of the photographs.

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Cite this article as: Chandravanshi SL, Rathore MK, Tirkey ER. Congenital combined eyelid imbrication and floppy eyelid syndrome: Case report and review of literature. Indian J Ophthalmol 2013;61:593-6.

Source of Support: Nil. **Conflict of Interest:** None declared.