

Congenital eyelid imbrication syndrome: A rare occurrence in Pakistan

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Abstract

Congenital eyelid imbrication syndrome (CEIS) is a rare condition presenting at birth and is characterised by overriding of the upper lid on the lower lid. It is due to longer upper lid, than the lower lid. Overriding leads to spontaneous eversion of the upper lids. In our patient, examination revealed canthal tendon laxity and hyperaemia of the tarsal conjunctiva. All the rest of the structures in the eyeball and adnexa were normal. Spontaneous eversion occurred in two weeks as the upper lid grew with time. No treatment was required.

Keywords: Congenital imbrication syndrome, Overriding upper lid, Hyperaemic conjunctiva, Floppy eyelids.

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Introduction

Congenital eyelid imbrication syndrome (CEIS) presents at birth with overriding long upper lid onto the lower lid which causes spontaneous eversion. For this, mostly babies are brought to an ophthalmologist soon after birth. There is also canthal tendon laxity which has been reported to improve between two days to two months. Various anatomical and physiological factors have been shown to be employed in its pathogenesis. A study has suggested the spasm of Orbicularis Ocularis to be the cause, which is suspected to occur as a result of irritation of upper tarsal conjunctiva by the lower lid. This sets up a vicious cycle.¹ It has been reported to occur as an isolated entity or along with Floppy eyelids and microphthalmia. It is mostly bilateral.^{1,2} Most cases that have been reported in Asia are associated with floppy eyelids.^{2,3} One case has also been reported from Hispanic origin.³ Regarding management, it has been postulated that topical lubricating agents like Carboxymethylcellulose reduce irritation, and topical Tobramycin provide prophylaxis against secondary infection.³ Other associations include microphthalmia, Down syndrome, Cat Eye syndrome, ptosis, and Ophthalmia neonatorum.⁴⁻⁶ According to one study it occurs as a result of involuntional changes leading to

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subsequent tightening of laxated canthal tendons and achievement of normal tone and size of the upper eyelids, in the first few weeks of life.⁷ The spontaneous eversion has been found to be directly related to the overriding of the lower lid by the upper lid.^{8,9} Kaur et al suggested thicker lipid layer and more mucin content of tear film in infants could add to the reversion of the condition.¹⁰ Various terms have been used to describe this condition. One study suggests that CEIS should be considered a variant of a spectrum of congenital ectropion.¹¹ One article has reported it as congenital bilateral upper lid eversion with severe chemosis.¹² Although spontaneous recovery is usually the rule, in some severe exceptions, medical and even surgical modalities of treatment have been described.¹³

We present this case to recall the diagnosis of CEIS which is although very rare but can present to a paediatric or paediatric ophthalmology clinic and could be mistreated due to lack of knowledge. In lieu of its course of spontaneous resolution it should only be observed and treated with only a lubricant. Parents must be counselled accordingly.

Case Report

An eight-day-old boy presented to the paediatric ophthalmology clinic at the Isra Post-graduate Institute of Ophthalmology, with spontaneous eversion of both upper lids mostly during crying (Figure: a), on December 2, 2022. On examination, marked conjunctival hyperaemia and bilateral canthal laxity were noted. Both the corneas were normal. Upon attempt to close, the upper lids were observed to override the lower lids by 2mm (Figure: b). No such presentation had been seen in any of the siblings. The upper lids could be inverted on attempted manual closure.

There were no signs of any other ocular pathology or



Figure: a) Initial presentation, eight days after birth. b) On two weeks follow-up.

systemic illness. It was diagnosed to be congenital imbrication syndrome. The parents were counselled regarding the spontaneous resolution of the condition and advised follow-up after two weeks. Application of lubricant ointment was advised four times a day. Upon follow-up after two weeks the upper lids had begun to stay inverted mostly and only partially everted laterally. After one further week the lids had completely inverted. There was no overriding of the lower lid by the upper lid.

No association was noted with microphthalmia, floppy eyelids, Down syndrome or any other condition as has been reported by other authors.

Informed consent was taken from the parents for the publication of this case report.

Discussion

CEIS is a rare condition. Limited data is available in literature. This is the first case reported from Pakistan. Limited number of such cases being reported could partly be because of the short course of the condition leading to rapid spontaneous recovery usually between two days to two months of birth.^{2,4,5} CEIS has been observed to be associated with floppy eyelids, microphthalmia, Down Syndrome, Cat Eye Syndrome, ptosis, and Ophthalmia Neonatorum.^{5,6-8}

The actual aetiology of CEIS is yet not clear. Possible mechanisms of resolution have been postulated by different authors. One of the article postulated that the whole eyelids were bulky and floppy and they underwent involuntional changes under the influence of unknown effects in the first week of life. This resulted in the tightening of laxated canthal tendons spontaneously. They negate the contribution of postnatal growth of the orbit in the normalisation of the eyelids.⁷ Others think that there could be the tightening of medial and lateral canthal tendons and involution of the upper lid with growth of bony orbit.⁹ Kaur et al suggested that the thicker lipid layer and more mucin content of tear film in infants as compared to adults enhances lubrication leading to spontaneous recovery.¹⁰

It was stated by Berger et al that the postnatal growth of the bony orbit does not bring spontaneous correction of eyelid imbrication because orbital growth in 1 week is not significant enough to correct 6 mm of overriding of eyelids.¹⁴

Conclusion

It is the first reported case from Pakistan. It must be reiterated that CEIS is self-resolving and no surgical intervention is required. Surgery is only required for Floppy eyelids which occurs in adults. Hence any such case should only be observed and no treatment options be attempted except for a lubricant, which might otherwise result in poor prognosis.

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Author Contribution:

NQ: Conception, drafting, revision, final approval and accountable for all aspects of the work.
SSB, FR: Data acquisition, revision, final approval and accountable for all aspects of the work.