Case Report

Congenital Combined Eyelid Imbrication with Congenital Floppy Eyelid Syndrome- A Rare Case Report

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Abstract *A healthy male term neonate born by a normal vaginal delivery was referred with severe bilateral eyelid edema and erythema at birth. On evaluation he was found to have eyelid imbrication with floppy eyelids bilaterally. The patient was managed conservatively and showed complete recovery of symptoms and signs over a period of ten days. This rare clinical entity can be a cause of concern to parents and clinicians but can be satisfactorily managed by conservative measures. This is the fourth case of congenital combined eyelid imbrication (CEI) with congenital floppy eyelid syndrome (CFES) reported worldwide, to our knowledge.*

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Introduction

Congenital eyelid imbrication syndrome (CEIS) is a rare clinical entity which is characterized by over-riding of upper eyelids over the lower eyelids.[1] It is observed at or shortly after birth and is generally bilateral in nature. It is essentially self-resolving with conservative[1] management, in contrast to adult-onset eyelid imbrication and floppy eyelids, which is acquired in nature and requires surgical intervention. In very rare cases, CEIS is associated with congenital floppy eyelid syndrome (CFES).[2] We report such a case of congenital combined eyelid imbrication with floppy eyelid syndrome, which was managed conservatively at our center. To our knowledge, this is the sixth case of congenital eyelid imbrication reported worldwide, and fourth reported case with an associated congenital floppy eyelid component till date. An informed consent has been obtained from the parents for submission of this case report.

Case Report

A three-hour old male neonate was referred for gradually increasing bilateral swelling and redness of the eyelids, with failure to open both eyes, since birth. The baby was born by a normal vaginal delivery at term and had no comorbidities or syndromic associations. There was no history of consanguinity among parents or in the family.

On evaluation, the baby was found to have bilateral bulky, edematous and erythematous eyelids, with over-riding of the upper eyelids over the lower eyelids by 6mm (Figure 1). The horizontal length of the upper eyelids was 23mm, while the mid-point of their vertical height was measured at 10mm. There was no spontaneous opening of the eyes. No other facial anomaly was noted. On applying gentle traction to open the eyes, the upper eyelids were found to evert spontaneously, and the tarsal conjunctiva was seen severely erythematous (Figure 2). The lower eyelids were folded inward bilaterally and when gently pulled down, they were also found to evert on minimal traction and the underlying conjunctiva was inflamed. There was mild mucoid discharge from the eyes. The globes were bilaterally normal on gross evaluation.[2]

Figure 1: *Patient photo at presentation showing bulky upper eyelids over-riding lower eyelids*

Figure 2: *Spontaneous eversion of eyelid with gentle traction on skin, with erythema of tarsal conjunctiva*

Figure 1: *Follow-up picture at four days post-partum with reduced bulkiness of upper eyelids*

Figure 4: *Follow-up picture at four days post-partum showing eversion of upper eyelids*

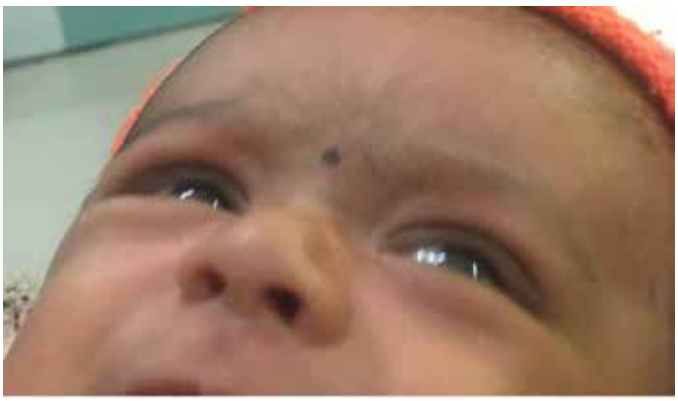

Figure 5: *Follow-up picture at ten days post-partum showing normal eye opening*

Based on the over-riding of upper eyelids over lower, eyelid laxity and eversion of eyelids, with inflammation of tarsal conjunctiva of both eyes, the patient was diagnosed as a case of congenital combined eyelid imbrication with congenital floppy iris syndrome.

Swabs were sent from upper and lower tarsal conjunctivae of both eyes for culture and were reported negative. The baby was managed conservatively with topical antibiotic (Tobramycin 0.3% eye drops) and lubricating agent (Carboxymethylcellulose 0.5% eye drops). The baby was routinely followed up and parents were counselled to alleviate anxiety regarding the clinical appearance. Over a period of ten days with conservative management, all symptoms and signs were resolved completely, and the baby achieved normal eye opening with reduction in eyelid laxity (Figures 3, 4, 5). Subsequent ophthalmic examination including fundoscopy was normal in both eyes.

Discussion

Congenital eyelid imbrication syndrome (CEIS) is a rare, self-resolving condition of unknown etiology, which may also be associated with congenital floppy eyelid syndrome (CFES). This essentially self-resolving entity can be a matter of anxiety among parents and healthcare staff at initial presentation, due to its clinical picture. The baby is characteristically found to have bulky, erythematous, over-riding upper eyelids, with mild to severe discharge from the eyes, at or shortly after birth. There is spontaneous lid eversion on gentle pull over the eyelid skin, and even on forceful crying in many cases.[3]

The etiopathogenesis of this condition is poorly understood and still under research. Many theories have been hypothesized to describe the course of events that occur from the onset to resolution of the clinical features. The over-riding of eyelids is said to be due to lid laxity and poor tone of the medial and lateral canthal tendons.[4] Irritation of upper tarsal conjunctiva by lower lid eyelashes may set up a vicious cycle of the irritation-spasm-over riding phenomenon.[3,5] The spontaneous eversion of eyelids is hypothesized to be due to persistent spasm of orbicularis oculi subsequent to irritation, which is probably aggravated as the eyelid skin is pulled to open the eyes, or when the baby is crying.[3,5] The management of these cases, hence, is based on alleviating irritation, thereby breaking the cycle. Topical lubricating agents like carboxymethylcellulose play a crucial role in reducing irritation, while topical antibiotics like tobramycin give prophylaxis against secondary infection. Sometimes there may be simultaneously existing ophthalmia neonatorum at presentation, and these cases need to be managed more aggressively depending on etiology. Hence, early reporting of cases and a reliable laboratory diagnostic facility are very important in satisfactory management of these cases. CEIS is said to spontaneously resolve due to tightening of medial and lateral canthal tendons with growth of bony orbit.[2,4] It has also been hypothesized that involution of upper eyelid structures in the first week after birth under yet unidentified influence may result in tightening of the canthal tendons and reduction in eyelid laxity.[6]

To this day, the etiology of this condition is largely unknown. CEIS has no proven syndromic associations. Congenital floppy eyelid syndrome has been associated with Down syndrome, however, our patient did not exhibit any features of Down syndrome or any other syndromic association.[7] Further research is needed to identify the etiopathogenesis of this condition.

It is [4]impeded to some extent, by the rare nature of the entity. However, it may be noted that many cases with very mild symptoms may go unreported, where the clinical picture may not be as dramatic and may resolve completely before it attracts the attention of the clinician.

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