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Case Report

Congenital Eyelid Embrication Syndrome Case Report

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Abstract

Congenital eyelid imbrication syndrome (CEIS) is an extremely rare, benign and usually self-limiting eyelid disorder. It is a bilateral condition that classically presents with a triad of upper eyelids overriding the lower eyelids, laxity of medial and lateral canthal tendon and last but not least tarsal conjunctival hyperemia.

We report a case of a healthy neonate that has congenital combined eyelid imbrication and floppy eyelid syndrome which resolved with conservative treatment in a week time. There are very few reported cases of this syndrome alone or in association with floppy eyelid syndrome.

Keywords: *Congenital Eyelid; Syndrome; CEIS; Hyperemia*

Introduction

Eyelid imbrication syndrome (EIS) is rare, there has been less than 10 cases are reported till date on PubMed search with keywords-congenital, eyelid imbrication. We hereby, report the 7th case of literature (1st Arabic origin) with bilateral congenital eyelid imbrication.

Case Presentation

A full-term newborn Emirate female, after uneventful normal vaginal delivery at 38 + 3 weeks, was referred to us 3 hours after delivery for inability to open the eyes that was observed immediately after birth. The parameters at birth were as follows: Birth weight 3919g, length 50 cm, head circumference 36cm and Apgar score Apgar score at 1 min was 8 and at 5 min was 9. She was the fourth child in the family. There was negative family history of ocular malformation, systemic or ocular syndromes or even consanguinity. An antenatal history revealed that Mother had gestational diabetes and was positive for group B streptococcus.

Ocular examination of child asleep showed elongated, bulky upper eyelids that overlapped on lower eyelids by 4 mm OU (Figure 1). Lower eyelids were not visible and covered by the upper eyelid along the entire length. The Vertical palpebral fissure in awakening state was 5 mm. Horizontal palpebral length was 18 mm while horizontal length of upper eyelid was 22 mm. The canthal tendons of both eyelids were symmetrically laxed, long and round in appearance. Both the upper eyelids were floppy on attempting a mechanical eyelid opening and showed spontaneous eversion even (Figure 2). Manual repositioning was required to put back the upper eyelid to normal position. Tarsal conjunctiva of both upper eyelids showed mild hyperemia and mild chemosis (Figure 3). The pre-tarsal region skin of lower eyelids was soggy and blanched secondary to mechanical pressure and lacrimation (Figure 4). Both corneas were clear and did not stain with

fluorescein ophthalmic drops. Digital intraocular pressure was within the normal range. Both eyes pupillary reactions are brisk. The rest of adnexal, anterior segment and dilated fundus examination were unremarkable. Conjunctival swab was taken and showed no growth. Condition was explained to the mother who was reassured. We advised using lubricants drops and follow up appointment for observation.

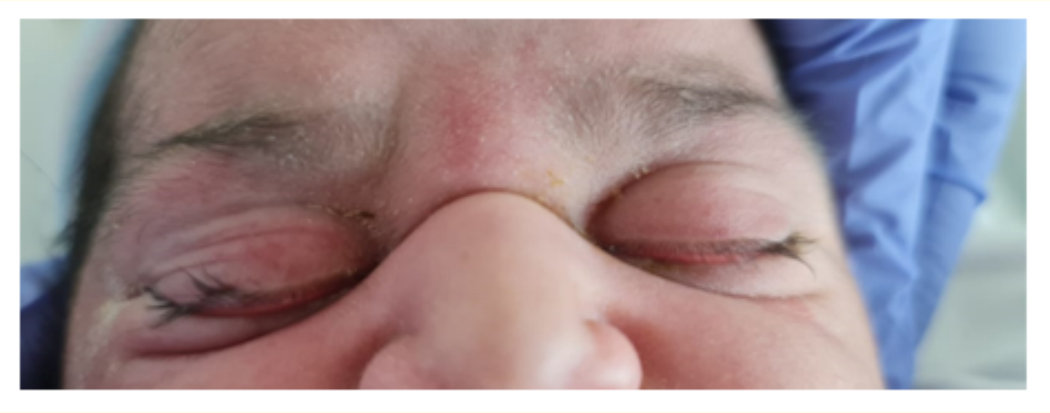

Figure 1: *Bilateral upper eyelid imbrication is noted clinically. The lower eyelid margin and eyelashes are not visible throughout the horizontal length.*

Figure 2: *A spontaneous upper eyelid eversion is noted on attempting a mechanical eyelid opening with spontaneous eversion even.*

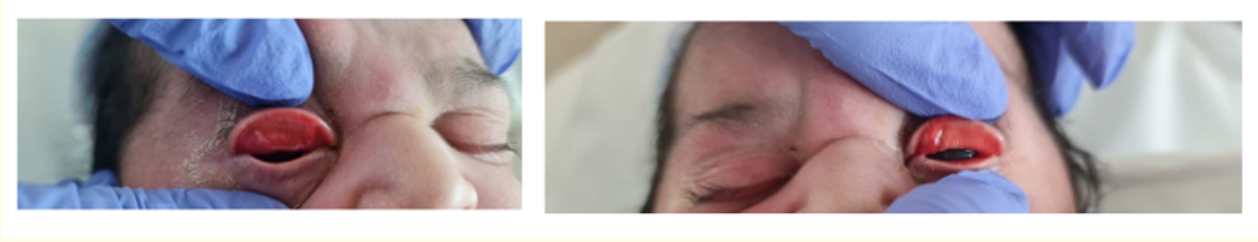

Figure 3: *Tarsal conjunctiva of both upper eyelids showed mild hyperemia and mild chemosis.*

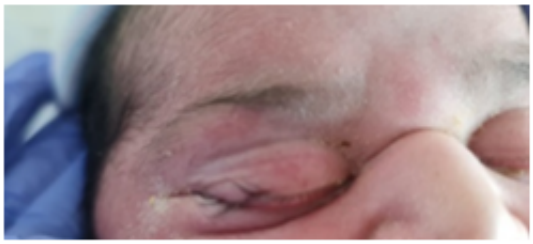

Figure 4: *The pre-tarsal region skin of lower eyelids was soggy and blanched secondary to mechanical pressure and lacrimation.*

Features	Rumelts., *et al.*	De silva., *et al.*	Odat., *et al.*	Chandravanshi., *et al.*	Kaur., *et al.*	Vempuluru., *et al.*	present
Age at presentation	3hr	48hr	➢ 24hr	16hr	1day	1day	3hr
Sex	F	M	M	F	M	F	F
Method of delivery	Vaginal	NM	Vaginal	Caesarian section	Caesarian section	Vaginal	Vaginal
Amount of overriding	OU 1.25mm	OU > 1mm	OD > OS OS 1-2mm	OU 6mm	OU4mm	NM	4mm
canthi	Laxed and longer	Laxed	Laxed	Laxed and longer	Laxed	NM	Laxed and longer
Spontaneous eversion of upper lid	Absent	Present	Present	Present	Present	Present	Present
Purulent discharge	NM	NM	Present	Present	Present	Present	Present
Papillary reaction	Absent	Few papillae	NM	Absent	Absent	Absent	Absent
Recovery	1week	2month	3week	1week	1week	8weeks	

Table 1

Discussion

Congenital EIS is a rare, benign idiopathic syndrome that affect both eyes and leads to eyelid malposition condition. It is self-limiting and can be accompanied with eyelid laxity that is characterized by the upper lid overriding the lower lid [1]. Usually associated with CFES [3,4]. Published literature on CEIS, have revealed an early presentation after birth with a mean of 22.75 hours (range 3 - 48 hours) and no sex predilection. The syndrome was also characterized with long lax canthi, spontaneous eversion of eyelids and spontaneous recovery. It can be accompanied with upper tarsal conjunctival hyperemia, secondary to the contact of the eyelashes with the lower lid [2-6]. Causing irritation and eventually a neuronal spasm of the orbicularis that increase in the override of the upper lid. CEIS is benign and does not need any surgical treatment [2,3]. Treatment basically aims to lubricate the ocular surface, leading to minimize the irritation caused by

the eyelashes. Researchers explained the self-limiting prognosis of the CEIS due to the growth of the orbit bones thus leading to spontaneous tightening of the canthal tendon, and therefore normal eyelid position [2,4].

Conclusion

We hereby, describe this rarely reported and possibly common condition in newly born infants. This is the first case of its kind reported in the Arab region population and the seventh reported case. Worldwide. As the condition resolves on its own without any sequela, that might be the reason of its scarce reporting in the scientific literature in our area and worldwide.

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