CONGENITAL EYELID EVERSION- A CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT: INTRODUCTION: Congenital eversion of the eyelids, a condition usually seen shortly after birth is a rare condition affecting one or both eyelids of the eye. Many etiologies have been described but no specific cause has been identified. We report a case of congenital eversion of eyelids which was managed conservatively with spontaneous correction seen in 6 days. **KEYMESSAGE:** Congenital eversion of the eyelids is a rare easily identifiable and potentially treatable condition. Decision of surgical intervention has to be taken with care and must be individualized for each case. Congenital eversion must be differentiated from congenital ectropion due to causes like ichthyosis as the treatment widely differs. All ophthalmologists need to be aware of this condition as it can be easily managed on an outpatient basis.
KEYWORDS: congenital, eversion, ectropion, eyelids.

INTRODUCTION: Congenital eversion of eyelids is a rare abnormality of the eyelids usually seen in otherwise normal babies early in life. This condition was first described as 'double eversion of the eyelids' by Adams in 1896.[1] It is also known as Congenital Eyelid Imbrication Syndrome. Various case reports have been published since then. We report one such case of unilateral congenital eversion of the eyelid with spontaneous recovery seen at the end of one week.

CASE HISTORY: A two day old baby was referred for examination with eversion of eyelids as shown in Image 1. Baby was born through a normal vaginal delivery. On examination child was crying and uncomfortable. Both upper and lower eyelids of both eyes were swollen and erythematous with crusting of the skin. Left eye upper lid was everted with exposure of the tarsal conjunctiva, with no signs of corneal exposure. Conjunctiva was congested and mild chemosis was noted. Cornea and the rest of the anterior and posterior segments were normal. Eyelid could be manually closed but would immediately Evert. Eversion was more when the child was crying.

Both the eyelids were patched after instilling hydroxypropyl methyl cellulose, ciprofloxacin eye ointment with forceful closure of the eyelids and lid taping. Patch was removed at 4 hourly interval for a period of 1 hour to prevent occlusion amblyopia. Hourly lubricating drops were prescribed and patching was continued. On day three (Image 2) congestion had come down and on forceful repositioning of the eyelids, the eyelids would remain in a normal position for around 3-5 minutes before everting. Upper eyelid was found to override the lower eyelid. Patching was discontinued and hourly lubricants and antibiotic ointment three times a day was used. At day six (Image 3 and 4) there was complete resolution of the eversion. At one month follow up there was no recurrence.

REVIEW OF LITEATURE: Congenital bilateral upper eyelid eversion is a rare condition. It is usually present at birth and bilateral. However unilateral cases and late onset beyond the first week of life have also been reported.[2,3] It is reported to occur predominantly in babies of dark skinned ethnic races, babies with Down's Syndrome[2], collodion baby syndrome[4] and Mongolism[3]. Most of the babies are otherwise normal. It is seen commonly after normal vaginal delivery but has also been seen after caesarean section.[5]

Eyelids are said to be spontaneously everting when they evert on gentle pulling of the skin of the upper eyelid towards the forehead, crying and yawning. The classical features have been described as a triad consisting of bilateral overriding of lower eyelids by the upper eyelids during sleep, bilateral medial and lateral canthal tendon laxity and tarsal conjunctival hyperemia.[5] Chandravanshi et al mentioned that the upper eyelid margins stained faintly with Rose Bengal suggesting drying of the exposed conjunctiva.[5] The chemosed and prolapsed conjunctiva probably protects the cornea and hence corneal problems are rare.[6] However, Al-Hussain et al reported a case of corneal perforation in a seven month old baby with upper eyelid eversion since birth with Down's Syndrome resistant to conservative management. The child required penetrating keratoplasty with lateral tarsorrhaphy and full thickness eyelid skin graft.[7]

Some case reports have categorized eyelid imbrication syndrome as a less severe form of floppy eyelid syndrome.[5] however we are of the opinion that both the entities are synonyms for the same condition and a larger case series would be helpful to establish or refute it.

ETIOPATHOGENESIS: Several factors have been implicated in the pathophysiology of development of congenital eyelid eversion. It was first proposed by Duke Elder that eyelids everted mechanically as the head traversed through the birth canal.[8] this was more common with face presentation of the foetus. Stillerman et al proposed that the facial trauma caused during labour led to impairment of venous drainage and periorbital oedema leading to eversion of the eyelids.[9]

Other intrinsic lid anatomical factors such as orbicularis oculi hypotonia, vertical shortening of the anterior lamella or vertical elongation of the posterior lamella of the eyelids with failure of fusion of the orbital septum with the levator aponeurosis, absence of an effective lateral canthal ligament and lateral elongation of the eyelids have also been implicated.[7]

Bentsi–Enchill proposed overlapping of the lower eyelid by the upper eyelid as a possible mechanism. This abnormality was seen in 22 of 150 new born babies in Africa in his study.[3] Spontaneous eversion is directly related to the amount of overriding of the upper lid on the lower lid.[10] With the overlapping lid and the face in a suitable position uterine contractions could possibly cause upper eyelid eversion and exposure of the conjunctiva which can cause irritation and blepharospasm. The blepharospasm will further prevent the reversion of the eyelid back to normal position. Venous congestion leading to chemosis will further aggravate the condition, setting up a vicious cycle. Congenital eversion though usually conservatively managed, in cases resistant to treatment, severe complications can be seen as the vicious cycle of venous stasis is exacerbated by delayed treatment. Early identification and intervention is of utmost importance.

TREATMENT: Many treatment methods have been described in literature. The conservative methods include eyelid repositioning, artificial tears, antibiotic ointments and regular eyelid patching. Spontaneous recovery is seen in most cases with conservative management as early as the third day.[11]

Eyelid repositioning includes mechanical repositioning which can be done by the mother followed by patching so that the lid stays in physiological state. However, patching must be done intermittently for 4 hours with a one hour interval to avoid the risk of occlusion amblyopia. The main goal of treatment is to prevent eyelid desiccation as prolonged drying will lead to failure of conservative management. This can be aided by the use of artificial tears. Chandravanshi et al demonstrated associated bacterial infection in congenital upper eyelid eversion leading to ineffective conservative management.[5] Ointment based antibiotics prevent and treat the infection with the added advantage of retaining moisture. Stern et al have described covering the eyelid with food wrap to prevent desiccation.[12] this method of treatment led to the inversion of eyelids in two to three weeks.

Isawumi et al advocated the use of hypertonic saline patches to reduce chemosis by osmosis.[13] W.A. Ibraheem has reported the use of gauze soaked in hypertonic saline to cover the chemosed and prolapsed conjunctiva for three hours once daily. Chemosis resolved significantly by day 3.[14] Watts and Dapling reported respiratory arrest during mechanical repositioning of the eyelid. Thus they emphasized the importance of vigilance towards autonomic effect of eyelid manipulations in neonates.[15]

Neonates with associated anatomical disorders or delayed treatment, severe degree of eversion, associated corneal complications may not respond to conservative management. Blechman et al reported a case of a day old baby who required surgery following unsuccessful conservative management due to non-attachment of orbital septum inferiorly to the tarsus.[16] Alvarez et al reported a case of failed conservative management at two months due to vertical elongation of eyelid in posterior aspect creating a skin deficiency.[17]

Many surgical techniques have been described for such cases. One such method is application of a fornix suture done by Bentsi–Enchill. In their study.[3] a double armed 3-0 silk suture was passed from the upper palpebral conjunctiva near the fornix to the skin just below the eyebrow and tied over a rubber bead. Antibiotic ointment and eye pad was applied with daily dressing. Eyelids reverted permanently within one to four days. Abiose has described the use of sub conjunctival sodium hyaluronate injection with resolution in 6 days.[18] Tarsoraphy has been described by Hopen.[19] Bentsi–Enchill also compared the results of 3 forms of treatment - lid suturing alone, sub conjunctival sodium hyaluronate injection alone and combined lid suturing and sub-conjunctival sodium hyaluronate injection. It was found that though all treatments worked well, in the combined approach of lid suturing and hyaluronate injection eyelids reverted to normal position in one to two days. It was proposed that this was superior to other treatments as conjunctival drying and infection is prevented. It would thus minimise the duration of hospital stay.[3] other methods include excision of redundant conjunctiva (Quoted by Stillerman ET al.[9]) or a full thickness upper lid skin graft.

Oluyemi Fasina has described the use of compression sutures in the management of eyelid eversion. He advocated passing 4-0 polypropylene sutures through the upper eyelid

margins after tucking in the chemotic conjunctiva and later sutured to the corresponding cheeks. Conjunctival protrusion significantly reduced in 36 hours with complete resolution of chemosis and reversion of the eyelid within a week.[20]

Many theories have been proposed for to describe the mechanism of spontaneous reversion. Rumelt et al suggested post-natal growth of the bony orbit to contribute to the spontaneous tightening of the previously lax canthal tendons resulting in correction of eyelid imbrication.[21] Chandravanshi et al however ascribed the resolution to involutional changes under the influence of unknown effect in the first week of life that led to tightening of lax canthal tendons and normalization of size and tone of the upper lid.[5]

Surgical intervention typically reduces the interval period of resolution of the eversion. However they are indicated only on failure of conservative management to break the callous cycle of strangulation and venous stasis which may lead to necrosis or exposure related complications. Ideally it is prudent to wait for spontaneous resolution with conservative management and resort to surgery only in cases with well documented failure of medical treatment. Nevertheless, cases with definite anatomical anomalies or severe cases with a tendency for complications need timely surgical intervention to prevent ocular morbidity as congenital eversion is 100% treatable.

Author	Cases	Laterality	Onset of		Modality of treatment	Resolution	Duration of treatment
			Disease	Treatment			
WA Ibraheem.[14]	1	Bilateral	6th days	6th days	Conser	11th day	5 days
Cingu et al.[11]	1	Bilateral	Birth	Birth	Conser	3rd day	3 days
Mazhar et al.[22]	1	Bilateral	Birth	7th day	Conser	2nd week	2 weeks
Silva et al.[23]	1	Bilateral	Birth	Birth	Nil	2nd month	2 months
Chandravanshi et al.[5]	1	Bilateral	Birth	Birth	Conser	7th day	7 days
Maheshwari et al.[6]	1	Bilateral	Birth	Birth	Conser	2nd week	2 weeks
Krishnappa et al.[24]	1	Bilateral	Birth	Birth	Conser	4th week	4 weeks
Al-Hussain et al.[7]	1	Bilateral	Birth	Birth	Surg	7th month	7 months
Rumelt et al.[21]	1	Bilateral	Birth	Birth	Conser	1st week	1 week
Oluyemi Fasina.[20]	1	Bilateral	Birth	Birth	Surg	1st week	1 week
Adeoti et al.[25]	3	Bilateral	Birth	Birth	Conser	8th day	8 days
		Unilateral	4th day	4th day	Conser	10th day	10 days
		Bilateral	Birth	Birth	Conser	10th day	10 days
Bentsi-Enchill.[3]	8	Bilateral	Birth – 6th day	At presentation	Lid Suture, SC Hyal, Lid suture + SC Hyal	Upto 2nd week	Upto 2nd week
	6	Unilateral					

Table 1: Demonstrates the various literature reported in relation to congenital eyelid eversion syndrome.

Conser-Conservative Management, Surg – Surgical Management after failure of Conservative Management, SC Hyal – Sub-conjunctival Hyaluronidase injection

CONCLUSION: Eversion of eyelids is a temporary problem which gets spontaneously corrected. Congenital eversion though an easily identifiable condition, has to be differentiated from congenital ectropion as children with ectropion require long term care while congenital eversion gets spontaneously resolved in days to weeks. Congenital eversion may rarely cause vision threatening complications but is mainly a cosmetic problem requiring close monitoring and early intervention to prevent complications.

LIST OF IMAGES:

A two day old baby with left eye congenital eversion of the upper eyelid. Right eye, upper lid overriding the lower lid – suggestive of imbrication.

Fig: 1

Same patient on day 3. Lids could be manually reverted to normal position. Overlap of lower lid by upper lid is seen.

Fig: 2

Image 3 & 4 – Day 6: complete spontaneous recovery with normal appearing conjunctiva

Fig: 3

Fig: 4

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