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Case Report

Ocular manifestation of a case of lamellar ichthyosis

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ABSTRACT

A baby full term born of LSCS to a otherwise healthy mother was referred for ophthalmology department. On ophthalmological torch light examination, the patient had equal upper and lower lid bilateral ectropion. There was missing eye lashes noted. The anterior segment evaluation was found to be within normal limit without evidence of corneal opacities. Lamellar ichthyosis otherwise termed as Collodion baby is a infrequent disease; and there is no well-known management protocol. The Patient was managed conservatively by the use of Hydroxy propyl methyl cellulose (2% w/v) eye ointment and lubricant (Carboxy methyl cellulose 0.5%) eye drops. Thus, surgical intervention was avoided and patient was managed medically on ointments and drops

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1. Introduction

Lamellar ichthyosis otherwise known as collodion baby, is a term for a baby who is born having skin enclosed in a tight shiny membrane that resembles a plastic wrap. It's congenital disorder of autosomal recessive type.¹ Seelingman in 1841 first described this condition and Hallopeau and Watelet² first introduced the term. Pérez in 1880 first gave the clinical description of this entity, which continues to be valid is "The baby's skin is replaced by a cornified substance of uniform texture, which gives the body a varnished appearance". The occurrence of this condition is 1case noted in 300,000 live births.³

Adams in 1986 firstly described the term congenital eversion of the eye lid and was termed "double congenital ectropion". This is usually a condition found bilaterally but unilateral cases have also been described. We are hereby reporting a rare clinical case of both lids' bilateral congenital ectropion, in an infant born with lamellar ichthyosis.

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2. Case Report

A full term baby born by LSCS, to a 23 year old healthy mother, having a non-consanguineous marriage, was brought to at our tertiary care centre at Neonatal intensive care unit (NICU) with signs of fissuring at places, dryness of skin, scaling. The infant was found having a congenital lamellar ichthyosis. The whole body on physical examination infant was seen to be enclosed with parchment-like membrane similar to a collodion and was shedding off from the entire body including the face. The mouth was continuously in an open position like a fish and There was slight eclabium, and. The patient was managed with appropriate fluids as well as electrolytes support along with prophylactic antibiotics cover. Emollients were given for skin softening and moistening in NICU.

The ophthalmology department was called upon for eye examination. On examination the patient was found having both upper lid and lower lid ectropion present bilaterally. Missing eyelashes were also noted as classical sign. The remaining of the anterior segment was within normal limit as There was no findings of watery or mucoid discharge,

opacities on cornea or keratitis noted on cornea. The patient was managed conservatively by the use of Hydroxy propyl methyl cellulose (2% w/v) eye ointment and lubricant (Carboxy methyl cellulose 0.5%) eye drops. Thus, there was no any requirement of surgical intervention and the infant was treated with appropriate drops and ointments.

3. Discussion

In this skin disorder ichthyosis which is found to have disproportionate drying of the skin and scales seen all over the body. The pathognomonic feature of this clinical entity is the sign of glistening brownish yellow membrane that encloses the entire body of the infant including face. Over a period of one month the membrane is usually replaced. Males are 2 times more likely to have the disease than females. Dehydration being the basic patho- physiology, adequate thermoregulation must be maintained by placing the baby in an environment which is heated with a bare minimum of at least forty to sixty % humidity. After initial stabilization during recuperating period, adequate and mandatory management for skin care should be given at the time of discharge to the family. The possibility of potential difficulties associated with this entity must be expected and minimized due to the impaired barrier function of the skin.

The chief priorities are diagnosing of this entity and urgent, proper care of the baby. During the neonatal period these infants are at higher risk of morbidity and mortality, because of the inadequate skin barrier function which increases the infant risk for higher water loss, thermoregulatory instability, skin toxicity and associated infection. In full term baby having such disease, they are at high risk for trans-epidermal water loss (TEWL) through skin and evaporation which is associated with heat loss. Flexor surface of the infant are at higher risk of being affected. There is also decreased sweating reported in such cases (10%).

Eye manifestations secondary to ectropion are in the form of corneal involvement as exposure keratitis, unilateral megalocornea, enlarged corneal nerve, other findings noted are absence of lacrimal puncta, blepharitis, absence of the meibomian gland, trichiasis, madarosis, and ectropion of both upper and lower lids.⁴ As per Maheshwari et al several possible mechanisms have been proposed but the main cause for eversion of the lids still remains obscure and associations have been recognized.⁵ Abnormalities noted such as vertical reduction in length of the anterior lamella or vertical extension of the posterior lamella of the eyelid, orbicularis hypotonia, birth trauma, and inability of the orbital septum to fuse with the Levator aponeurosis, ineffective lateral canthal ligament and lateral extension of the eyelid, have all been implicated as reasonable mechanical factors contributing.⁶ Stasis of Venous flow during labour may be associated with marked chemosis and prolapse of the conjunctiva, resulting in eversion of

the eyelids. After eversion, spasm of orbicularis becomes a sphincter that leads to a vicious cycle of conjunctival strangulation and oedema, secondary to venous stasis.⁷ The cornea is hence saved by chemosis and prolapsed conjunctiva and the complications associated with cornea is rarely seen.

There are certain studies which pointed that such entity managed conservatively by local use of clobetasol.⁸ In the early period of life in majority of the cases Ectropion in lamellar ichthyosis (collodion baby) have been found to respond to conservative management. Congenital eyelid eversion can be treated conservatively. To prevent desiccation of the exposed conjunctiva and the allow spontaneous inversion of the lid is the goal of management. tarsorrhaphy done for Temporary period, hyaluronic acid given sub conjunctival, sutures in the fornix and entire thickness graft of skin to the upper lid⁹ are the Surgical treatment options.

The future prognosis of these children is very good if proper care from dermatology, ophthalmology and paediatric department is taken. Extremely good results due to A comprehensive early management in such cases were seen and we are able to avoid surgical intervention.

4. Conclusion

Congenital ectropion found bilaterally is the most frequently found ocular manifestation in the infant with lamellar ichthyosis which is best managed conservatively. Proper patching of the eye with appropriate ointment and topical application of lubricating eye drops are the best initial line of treatment. Surgical intervention is planned only if conservative management does not give desired results.

5. Conflict of Interest

The authors declare no relevant conflict of interest with respect to research, authorship and or publication of this article

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