

Congenital Bilateral Ectropion in Collodion Baby: A Rare Case Report

Aruna K.R. Gupta, Kashyap Patel, Yashvi Nathwani, Nikunj Amin, Kishan Makwana

Dept of Ophthalmology, C.U. Shah Medical College & Hospital, Surendranagar, Gujarat, India

Abstract

A baby born with normal full term vaginal delivery to a healthy mother aged 30 years was admitted to Neonatal intensive care unit at our tertiary care centre. On ophthalmological examination, the patient had bilateral ectropion of both the upper and lower eyelids. The eyelashes of the lower eyelids were missing. There was no evidence of discharge, corneal opacities or keratitis. No other ocular abnormality was found. Collodion baby is a rare disease; and there is no established management protocol. We managed the patient conservatively by the use of 0.05 % Clobetasol ointment for local application over the eyelids and lubricant (Carboxy methyl cellulose 1%) eye drops. Thus, the case was managed successfully with no requirement for surgical intervention.

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Introduction

Collodion baby, also known as Lamellar Ichthyosis, is a descriptive term for an infant who is born encased in a tight shiny membrane that resembles a plastic wrap. It is a form of congenital ichthyosis and is an autosomal recessive disorder.¹ This condition was first described by Seelingman in 1841 and the term was first introduced by Hallopeau and Watelet.² The first clinical description of collodion membrane by Pérez in 1880, 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 incidence of this condition is 1 in 300,000 live births.³

Congenital eversion of the upper eyelid in a collodion baby was first described by Adams in 1986 and was termed "double congenital ectropion". The condition is usually bilateral but unilateral cases have also been reported. We are hereby reporting a rare case of bilateral congenital ectropion, in a baby born with lamellar ichthyosis.

Case Report

A baby born at normal full term, by vaginal delivery, to a healthy mother aged 30 years, of a non-consanguineous marriage, was admitted to Neonatal intensive care unit (NICU) at our tertiary care center with signs of dryness of skin, scaling, fissuring at places and severe itching. The baby was diagnosed to have congenital lamellar ichthyosis (Figure 1a). On physical examination the whole body was found to be covered with parchment-like membrane resembling a collodion and was peeling off from the entire body including the face. There was slight eclabium, and the mouth was constantly in an open position like a fish. The patient was managed in the NICU with appropriate fluid and electrolytes along with prophylactic antibiotics. Emollients were advised for skin softening and moistening.

The patient was referred to the ophthalmology department. On examination, the patient had bilateral ectropion of both the upper and lower eyelids. The eyelashes on the lower eyelids were missing. There was no evidence of discharge, corneal opacities or keratitis. The rest of the anterior segment was normal (Figure 1b). We managed the patient

conservatively by the use of clobetasol 0.05% ointment for local application over the eyelids twice a day and hourly instillation of lubricants (carboxymethylcellulose 1.0%) in the eye. Wet saline gauze was lightly placed over the face and totally covering the eyes.

The patient was discharged after one week. Regular follow up to the patient was advised and any progression was noted. The shining membrane present all over the body cracked and peeled off over the course of several weeks. Ectropion and eclabium were also resolved at the end of 3 months (Figure 1c).

Discussion

Ichthyosis is a skin condition characterized by excessive drying of the skin and scales over the body. The characteristic feature of the disease is the shinning brownish yellow membrane that covers the entire body of the baby. The membrane is usually replaced over a period of one month. The disease is reported to be two times more common in males than in females. Since the basic pathology is dehydration, proper thermoregulation is achieved by placing the infant in a heated isolette with a minimum of 40 to 60% humidity. After the initial stabilization and convalescent period, proper guidance for skin care should be given at the time of discharge to the family. Due to the impaired barrier function of the skin, the possibility of potential complications associated with this condition should be anticipated and minimized.

Identification of this condition and immediate, proper care of the infant are the chief priorities. Morbidity and mortality in these children during the neonatal period is high, because of the impaired skin barrier function which places the infant at risk for increased water loss, thermal instability, percutaneous toxicity and infection. In a term collodion baby, the infant is at risk for trans-epidermal water loss (TEWL) and associated heat loss that accompanies evaporation. Flexor aspects of the body are more commonly affected. The other condition reported is diminished sweating (10%).

Ocular manifestations may be in the form of exposure keratitis secondary to ectropion, unilateral megalocornea,



Figure 1a: Congenital lamellar ichthyosis



Figure 1b: Bilateral ectropion noted at birth



Figure 1c: Improvement in ectropion after 3 months

enlarged corneal nerve, blepharitis, absence of the meibomian gland, trichiasis, madarosis, absence of lacrimal puncta and ectropion of both upper and lower lids.⁴ According to Maheshwari et al the underlying cause for eversion remains obscure and several possible mechanisms have been proposed and associations have been recognized.⁵ Abnormalities such as orbicularis hypotonia, birth trauma, vertical shortening of the anterior lamella or vertical elongation of the posterior lamella of the eyelid and failure of the orbital septum to fuse with the Levator aponeurosis, absence of effective lateral canthal ligament and lateral elongation of the eyelid, have all been implicated as possible patho-physiological factors.⁶ Venous stasis during delivery may cause marked chemosis and prolapse of the conjunctiva, causing eversion of the eyelids.⁷ Once everted, orbicularis spasm may act as sphincter that leads to a vicious cycle of conjunctival strangulation and edema, secondary to venous stasis.⁸ The chemosed conjunctiva protects the cornea from exposure and hence, corneal complications are rare.

There is evidence which suggested that ectropion in collodion babies can be managed conservatively by local application of clobetasol.⁹ Ectropion in lamellar ichthyosis (collodion baby) may respond to conservative management in the early period of life in about half of the cases. Congenital eyelid eversion can be treated conservatively. The goal of management is to prevent desiccation of the exposed conjunctiva and the allow spontaneous inversion of the lid. Surgical treatment options include temporary tarsorrhaphy, subconjunctival injection of hyaluronic acid, fornix sutures and full thickness skin graft to the upper lid.¹⁰

The future prognosis of these children is very good if proper care from dermatology, ophthalmology and pediatric department is taken. A comprehensive early management in such cases has shown extremely good results and we are able to avoid surgical intervention.

Conclusion

Bilateral ectropion is the most common ocular finding in cases of lamellar ichthyosis which is initially best managed conservatively. Topical lubricants and proper patching with regular follows ups is the initial line of management. Surgical intervention is planned only if conservative management does not give desired results.

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Address for correspondence

Aruna K.R. Gupta MBBS, DNB, MNAMS
Dept of Ophthalmology, C.U. Shah
Medical College & Hospital, Dhudrej Road,
Surendranagar, Gujarat - 363001, India
Email id: arunagupta.eye@gmail.com



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