Acta Scientific Ophthalmology (ISSN: 2582-3191)

Volume 4 Issue 9 September 2021

Case Report

A Case Report of Lamellar Ichthyosis

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Received: August 09, 2021

Published: August 28, 2021

Abstract

Ichthyosis is an inherited skin disorder (also called disorders of keratinization) is characterized by hyperproliferation of cells and thus an abnormal physical barrier and manifests as dry, scaly and thickened skin. Lamellar ichthyosis (LI) is one of the subtypes and has an autosomal recessive inheritance. It is a rare condition where babies present with shedding of skin and is usually generalized. They are also called as "collodion babies" as they are said to have a shiny armor of shed skin [1]. Here we present a case of a newborn baby diagnosed with LI and bilateral ectropion which spontaneously reduced after topical treatment such as lubricants and antibiotics were started. We are reporting this case as LI is considered to be a rare inherited disorder and the bilateral ectropion was found to reduce spontaneously without any invasive interventions.

Keywords: Lamellar Ichthyosis; Bilateral Ectropion

Introduction

Ichthyosis are a group of inherited disorder which is characterized by thickening and scaling of the skin. It occurs as a result of abnormal cornification. There are subtypes of ichthyosis; autosomal dominant- ichthyosis vulgaris, x-linked ichthyosis, autosomal recessive- lamellar, and congenital form-ichthyosiform erythroderma [4].

Lamellar ichthyosis is a rare autosomal recessive disorder affecting 1 in 300.000 people [4]. The most common ocular manifestation of the eyes reported is cicatricial ectropion. This will further lead to corneal exposure and thus exposure keratopathy.

Conservative management includes methyl cellulose eye drops, antibiotics to prevent superadded infection. In certain cases, multimodal approach may be required i.e. surgical along with medical treatment. Surgical treatment includes retroauricular full/split thickness skin graft for the upper eyelids and cheek transpositions have been done for lower eyelids [4].

Case Report

A newborn baby with polygonal shaped, brown, adherent scales all over the body who was diagnosed with lamellar ichthyosis (LI) (Figure 1) was referred from the pediatrics department to ophthalmology for bilateral outward turning of upper eyelids.

Baby was born to parents with no history of consanguineous marriage among them. It was a full term normal vaginal delivery at 39 weeks of gestation. Apart from the dermatological and ocular manifestations (in the form of ectropion), there were no other significant findings. The family history was also found be normal.

Figure 1: A newborn baby with polygonal shaped, brown, adherent scales all over the body- a case of lamellar ichthyosis (LI).

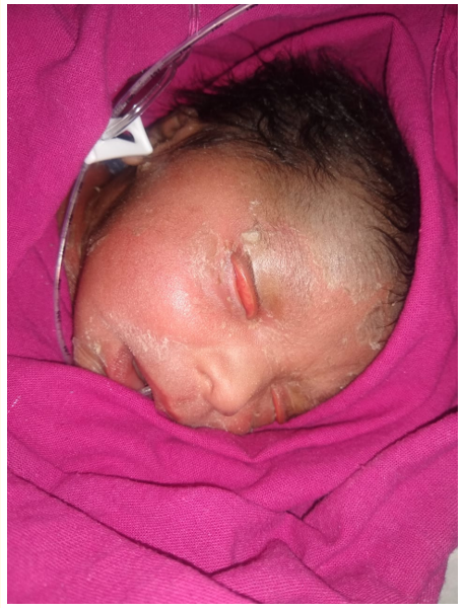

Figure 2: Bilateral upper eyelid ectropion.

On general examination the baby was found to have large areas of scaly skin that was in the form of thin membranes and was tightly adherent to the underlying skin in the center.

On ocular examination, both upper eyelids showed ectropion however, lower eyelids were found to be normal (Figure 2). On retraction of both eyelids, cornea, anterior chamber depth was normal and pupils were found to be briskly reacting to light. As a part of the complete eye examination, fundoscopy was performed and was found to be normal.

As a part of the treatment, lubricant eyedrops coupled with a topical antibiotic drop (to prevent super-added infection) was prescribed. The baby was monitored everyday and was found that the ectropion improved significantly within the first 24 hours. Complete and spontaneous resolution of ectropion was seen in 6 days (Figure 3).

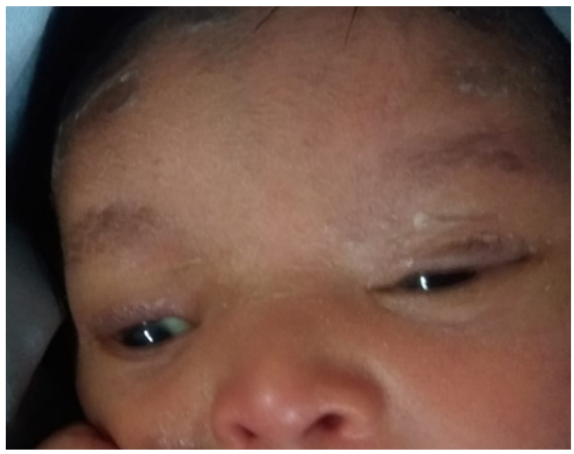

Figure 3: Spontaneous improvement in ectropion as seen on day 6.

Discussion and Conclusion

Lamellar ichthyosis (LI) is a rare clinical manifestation (seen in 1:300,000 births) [4]. Infants born with this condition most commonly are born with an adherent, clear sheath covering their skin, called as collodion membranes which is usually shed during the initial few weeks and thus leaves behind a scaly skin.

Ocular manifestations include exposure keratitis (secondary to ectropion), unilateral megalocornea, enlarged corneal nerves, blepharitis, deficit of the meibomian glands, trichiasis, madarosis, and absence of lacrimal puncta [5].

This case report highlights the ocular manifestations of Lamellar ichthyosis mainly bilateral ectropion. In severe cases, it presents as cicatricial ectropion. It is observed that there is gradually progressive and abnormal cornification of the eyelid skin thus resulting in cicatrization and shortening of anterior lamella which eventually causes ectropion of eyelids. Exposure keratopathy follows suit due to the ectropion [2].

Emollients and oral retinoids are usually prescribed. Emollients act as moisturizers and retinoids are used as they improve healing by decreasing epidermal proliferation. It is important to identify and treat sight threatening conditions such as ectropion (especially when it is caused due to cicatrization and thus leading to lagophthalmos) as early as possible to prevent complications like corneal melting [3]. Ectropion can usually be treated conservatively as was done in this case. However, severe cases require interventional/surgical correction like full thickness post auricular or split thickness skin graft from the thigh for the upper eyelids with 0.1% isotretinoin applied locally post procedure at both donor and recipient sites [2,6]. Lower eyelids if affected can be managed by cheek transposition grafts and lateral tarsal strip procedure [4,7].

It becomes crucial to adopt a multimodality approach in these cases. However, in our patient, given the severity of ectropion, application of emollients with topical antibiotics (Tobramycin eye drops 0.3%) was advised. Patient's mother was then counselled thoroughly and was asked to follow up on regular intervals.

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Volume 4 Issue 9 September 2021