

CASE REPORT

Lamellar Ichthyosis with Bilateral Ectropion

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ABSTRACT

Lamellar ichthyosis is a rare congenital disorder with ocular manifestation resulting from the cicatrization of anterior lamella of eyelids. Early diagnosis and management of ectropion can prevent the most severe complications such as severe dry eyes, corneal perforation, and possible loss of the eye. This case report highlights a case of lamellar ichthyosis with mild ectropion which was managed conservatively.

Keywords: Ectropion, ichthyosis, lamellar ichthyosis

INTRODUCTION

Ichthyosis is a heterogeneous group of dermatoses characterized by thickening, scaling, and fissuring of skin. Ichthyotic disorders are usually inherited, but sometimes may be acquired. It is divided into congenital and acquired types.^[1] Lamellar ichthyosis is an extremely rare condition with autosomal recessive inheritance, with a prevalence of 1 in 3,00,000.^[2] It usually presents at birth, and is associated with diffuse large, thick, brown pasted (platelike) scales. It has generalized lesions, accentuated on lower extremities and flexural areas. The ocular abnormality is mostly cicatricial ectropion, which can lead to exposure keratopathy and corneal perforation, when severe.^[3,4] Sometimes corneal exposure and dryness are very severe. Bilateral corneal perforation may be observed.^[5]

CASE REPORT

A 9-month-old female child was presented to the Pediatric Department of a Rural Based Tertiary

Level Referral Centre, Uttar Pradesh Rural Institute of Medical Sciences and Research, with a history of fish-like scales all over the body associated with watering in both eyes since birth. She also had a history of collision membrane at birth. She was born to a 24-year-old female with full term, normal vaginal delivery at a government hospital, in a rural area of Northern India. She was the third child of her parents. She had low birth weight of 2 kg for which she was kept in Neonatal Intensive Care Unit during perinatal period. The marriage of parents was consanguineous in nature.

On examination, the patient had generalized involvement of the whole body including face by fish-like scales, which were associated with minimal erythema [Figure 1]. She was referred for the ophthalmology consultation for the complaints of persistent watering in both eyes. On ophthalmological evaluation, the patient had grade I bilateral lower lid ectropion [Figure 1a]. Puncti were not apposed in both eyes. Lacus lacrimalis was present. There was no conjunctival congestion, and any signs suggestive of intraocular disease. There was no lagophthalmos in both eyes. Mild corneal dryness was observed in clinical evaluation but cornea was clear and transparent in both

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www.nigerianjournalofophthalmology.com

DOI:

10.4103/0189-9171.170996

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How to cite this article: Jain G, Jain VK, Sharma R, Sharma IK, Verma GK. Lamellar ichthyosis with bilateral ectropion. Niger J Ophthalmol 2015;23:65-6.

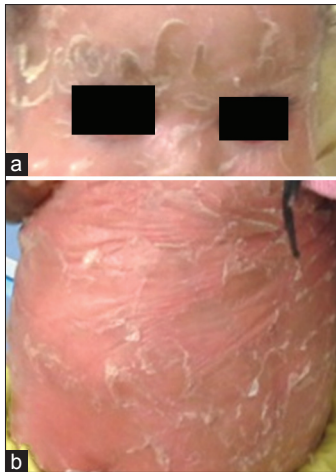


Figure 1: Clinical photograph of a 9-month-old child with bilateral lower lid ectropion and fish-like scales over the face (a) and abdomen (b)

eyes without any signs of exposure keratopathy. Based on the above clinical features, the diagnosis of lamellar ichthyosis with bilateral ectropion was made. As the child was having mild case of ectropion, she was treated with frequent lubricants (carboxymethylcellulose 0.5%) and topical antibiotics (moxifloxacin 0.5%) to prevent exposure keratopathy. The child was treated for the skin condition with hydration, lubricating, and keratolytic agents. She was kept under regular follow-up. After 2 weeks of starting treatment, child showed an improvement in skin condition as well as ectropion. At her last visit (6 months), the scaling and cicatrization was decreased, ectropion was also corrected, and watering got resolved. The child is currently under follow-up and advised for routine ophthalmic examination to observe for any worsening of disease and associated corneal abnormalities such as exposure keratopathy and corneal perforation.

DISCUSSION

Ichthyosis is a dermatological disorder characterized by hyperkeratosis and excessive dry skin. At birth, child usually presents as a collodion (lacquered) baby ensheathed in a membrane. After shedding of membrane, fish-like scales will manifest. The patient develops large, thick, brown pasted scales (platelike) which will persist for life. Flexures may show continuous linear rippling. Erythema is minimal or absent; when present, it is intense on the face. Dental abnormality is also reported in ichthyosis.^[6]

Lamellar ichthyosis is associated with ophthalmic involvement. Eye manifestations include ectropion, blpharitis, dry eyes, exposure keratopathy, corneal perforation, trichiasis, and madarosis.^[7,8] Ectropion

generally results from shortening of anterior lamella and termed as cicatricial ectropion. Cicatricial ectropion in mild form can be managed by conservative measures such as frequent lubrication. The severe form of cicatricial ectropion can be corrected by surgery.^[9] For the correction of cicatricial ectropion, anterior lamellar reconstruction with free skin grafts or mucous membrane graft can be done.^[10]

In our case, the ectropion was in milder form, so could be managed conservatively. Early diagnosis and treatment of ectropion with ichthyosis can prevent the later major ophthalmic complications such as corneal perforation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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