

## Combined medical and surgical management for cicatricial ectropion in lamellar ichthyosis: A report of three cases

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Ichthyosis is a rare inherited skin disorder characterized by abnormal keratinization of the epidermis. Cicatricial ectropion is the most common ophthalmic feature of congenital ichthyosis. Progressive subepithelial cicatrization and abnormal cornification of eyelid skin cause progressive ectropion in both eyelids, leading to lagophthalmos and corneal exposure. Surgical correction of cicatricial ectropion in these cases is challenging with unsatisfactory results. Proper processing of the donor and recipient site with lubricants and topical retinoids before surgery makes grafting easier and its survival better. We present three cases of lamellar ichthyosis with cicatricial ectropion managed with combined preoperative topical therapy followed by surgery. All patients had extremely good surgical outcomes, with none of them requiring repeat surgery.

**Key words:** Cicatricial ectropion, lamellar ichthyosis, skin grafting

Ichthyosis is a rare condition characterized by increased formation of epidermal scales and extreme dryness of the skin

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<b>Quick Response Code:</b>	<b>Website:</b> www.ijo.in
	<b>DOI:</b> 10.4103/ijo.IJO_855_20

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Received: 05-Apr-2020

Revision: 24-May-2020

Accepted: 19-Jul-2020

Published: 26-Oct-2020

throughout the body.<sup>[1]</sup> There is progressive and abnormal cornification of the eyelid skin, resulting in subepithelial cicatrization and progressive ectropion of eyelids, restrictive lagophthalmos, and exposure keratopathy.<sup>[1]</sup> Management of cicatricial ectropion in lamellar ichthyosis is primarily surgical and challenging because of its difficulty in harvesting the graft, its poor survival, and recurrence.<sup>[2]</sup> We present a small case series of three patients wherein both the donor and recipient sites were pretreated medically with lubrication and 0.1% isotretinoin ointment for a short duration followed by surgery. To the best of authors' knowledge, there is no report of such a treatment protocol in the literature.

### Case Reports

#### Case 1

A 4-month-old male baby with lamellar ichthyosis presented to us with complaints of inability to close both eyes since birth. On examination, skin all over the body was dry and scaly, and he had cicatricial ectropion of both upper lids [Fig. 1a]. The patient was planned for split-thickness skin graft (SSG) from the thigh. He was started on a 2-week course of liberal liquid paraffin all over the body and 0.1% isotretinoin ointment locally around the donor and recipient site to prepare both the areas for grafting. The donor and recipient sites had improved after 2 weeks of treatment, and the patient underwent ectropion correction with SSG in both the eyes. Postoperatively ectropion had corrected well, and there was no lagophthalmos [Fig. 1b]. The patient was advised to continue with liquid paraffin and ocular lubricants and was kept under regular follow-up. He was doing well at three years of follow-up [Fig. 1c].

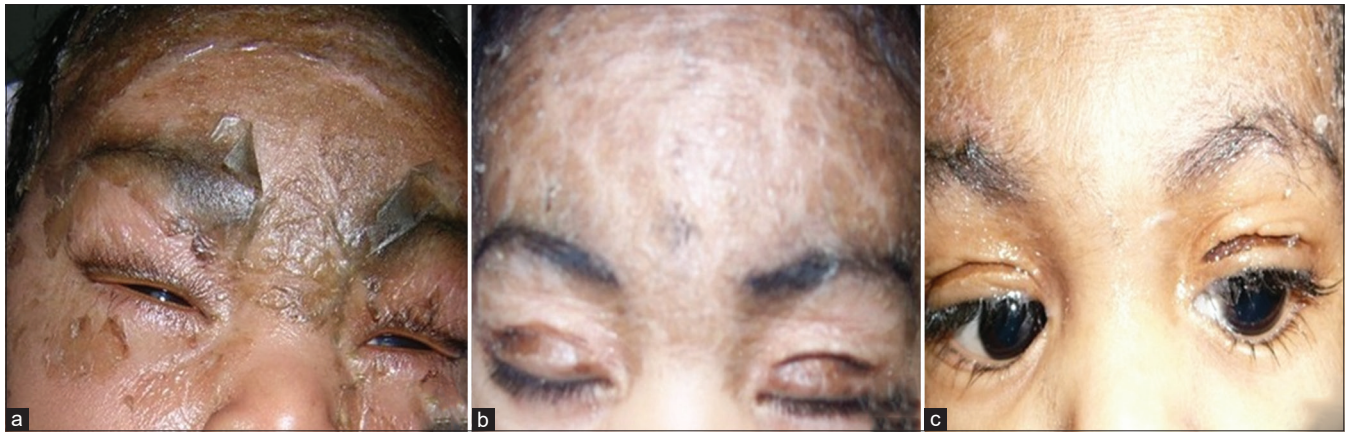
#### Case 2

A 6-year-old male child with lamellar ichthyosis presented with cicatricial ectropion of lower lids of both the eyes causing lagophthalmos [Fig. 2a]. An FTSG for the lower lid of both the eyes was planned. He was then started on liquid paraffin liberally and 0.1% isotretinoin ointment locally around the donor and recipient sites for 2 weeks. Lower lid ectropion correction was done with FTSG after 2 weeks of therapy. Postoperatively the ectropion had corrected well with minimal

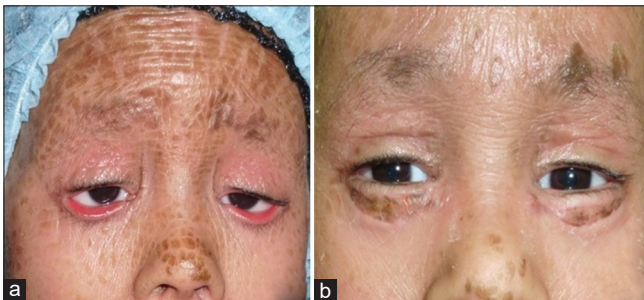
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**Cite this article as:** Subramanian N, Nivean PD, Alam MS. Combined medical and surgical management for cicatricial ectropion in lamellar ichthyosis: A report of three cases. Indian J Ophthalmol 2020;68:2615-7.



**Figure 1:** (a) Preoperative external photo of the infant showing cicatricial ectropion of both upper lid. Scales all over the face can be seen. (b) Postoperative photograph showing good graft uptake with correction of ectropion. (c) Postoperative photograph showing good anterior lamella of both the upper lids at 3 years of follow-up



**Figure 2:** (a) Preoperative external photograph of the young boy with cicatricial ectropion of both the lower lids. (b) Postoperative photograph of the patient showing correction of lower lid ectropion. Notice the improvement in the condition of upper lids, forehead, and nose because of medical therapy



**Figure 3:** (a) Preoperative external photograph of an adult lady with ichthyosis cicatricial ectropion of all four lids. (b) Postoperative photograph showing good graft uptake in all the four lids and good correction of ectropion

residual lagophthalmos [Fig. 2b]. The patient was kept on regular ocular lubricants and topical emollients. He was doing well at five years of follow-up.

### Case 3

A 50-year-old female presented to us with complaints of defective vision in both the eyes for the past 6 months. She had an age-related cataract in both the eyes and cicatricial ectropion of all four lids [Fig. 3a]. The patient was advised to use liquid paraffin all over the body and 0.1% isotretinoin ointment over the donor and recipient area for 2 weeks. The skin condition improved after 2 weeks, and she underwent

ectropion correction with SSG [Fig. 3b]. A suture tarsorrhaphy was done, which was removed after 24 h. The graft uptake was good, and the ectropion was corrected well. She underwent cataract surgery in both her eyes after 2 months. The condition of the eyelids was good without any ectropion at three years of follow-up.

### Surgical technique

The incision is made in the upper eyelid starting from above the upper punctum and laterally ending above the lateral canthus. Preferably the incision should be at the supratarsal fold. The incision is deepened dividing the fibrous tissue. Orbicularis is split and not divided. Two sutures are taken through the lid margin and taped to the cheek to get the overcorrection of the release. After establishing good hemostasis, the impression of the raw area is taken to harvest the graft. Similar surgical steps are followed for the lower eyelid. For the full-thickness skin graft (FTSG), the groin area is chosen. After being placed over the recipient site, the graft is sutured to the edge of the defect with ends of the sutures left long. These sutures are used to tie over a bolster. The idea is to prevent movement between the graft and the bed. The sutures are removed after five days to inspect the graft in the case of SSG and after seven days in the case of FTSG. After 2 weeks, a silicone gel sheet is used to cover the skin graft at least 6 h daily. Moisturizers are continuously applied postoperatively over the whole body and the recipient site.

### Discussion

Lamellar ichthyosis is an infrequent clinical entity worldwide (1:300,000 births).<sup>[1]</sup> Infants with this condition are typically born with a tight, clear sheath covering their skin, referred to as collodion membranes. This membrane is usually shed during the first few weeks of life, leaving behind diffuse dry, scaly skin for the rest of life.

Ocular manifestations include exposure keratitis secondary to ectropion, unilateral megalocornea, enlarged corneal nerves, blepharitis, absence of the meibomian glands, trichiasis, madarosis, and absence of lacrimal puncta.<sup>[1,3]</sup>

Surgical correction is the mainstay of management in cicatricial ectropion resulting from lamellar ichthyosis; however, successful management has been reported using

topical retinoids in few cases.<sup>[4,5]</sup> The main drawback of medical management is its long term need and recurrence of ectropion after cessation of medications. In the case series by Hanson *et al.*, where tazarotene skin cream was used for congenital ectropion, a successful outcome was seen in only two out of five cases.<sup>[5]</sup>

Surgical management is challenging mainly for two reasons. Firstly the harvest of graft is difficult because of the thin papery skin, and secondly, these kinds of grafts are notorious for poor uptake and survival.<sup>[2]</sup> In our series, we took FTSG or SSG based on the size required. FTSG was used mostly for lower eyelids, as can be seen in case number 2. SSG was used when the area was larger as-is for upper eyelids and in cases where all the four eyelids had to be resurfaced (Cases 1 and 3). The donor site for SSG in cases 1 and 3 was thigh while it was groin for FTSG. Post auricular site was avoided as there are high chances of secondary contracture in these areas.

We propose preparation of both the donor and recipient site with liberal lubrication and application of retinoid ointment before surgery for a considerable duration of at least 2 weeks. For this, we used liquid paraffin and 0.1% isotretinoin skin ointment. The lubrication with liquid paraffin moistens the skin while the retinoid decreases epidermal proliferation, inflammation, and promotes healing.<sup>[4,5]</sup> Once the donor area is ready for harvesting, which would be obvious by its appearance (soft and pink), the patient can be taken up for surgery.

The lubrication and retinoid also decrease the amount of ectropion by reversing skin cornification. Though we got the desired result with 2 weeks of retinoid application, the duration can be further increased depending upon the response.

Some novel surgical techniques in the form of using prepuccial skin and maternal skin graft have been reported, but again they are single case reports.<sup>[6,7]</sup>

Even the skin graft develops flaking as Ichthyosis is a disorder of keratinization and cornification; hence, the graft also needs frequent application of emollients. Secondary contracture of the skin graft can occur. We recommend a silicone gel sheet application at least for 3 months to reduce postoperative contracture. Ichthyosis does improve as the child grows, but it is necessary to use moisturizers throughout life. Any oil or liquid paraffin is useful.

## Conclusion

Combined medical and surgical management, i.e., preoperative skin lubrication and the application of topical retinoid ointment followed by surgical correction of ectropion, is an attractive option for cases of lamellar ichthyosis. It not only makes graft harvesting easier but also enhances their uptake and survival.

## 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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