

Cicatricial upper and lower eyelid ectropion in an ichthyosis patient. Surgical correction.

Ozlen Rodop Ozgur¹, Levent Akcay², Nesrin Tutas³, Yelda Ozkurt⁴

1. Dr. Lutfi Kirdar Kartal Training And Research Hospital, 1st Eye Clinic, Kartal, Istanbul, Turkey.

2. World Eye Hospital, Altinuzade, Uskudar, Turkey.

3. Kadirli State Hospital, Eye Clinic, Osmaniye, Turkey.

4. Fatih Sultan Mehmet Training And Research Hospital, Eye Clinic, Bostanci, Turkey.

Corresponding author:

Ozlen Rodop Ozgur, MD

Basibuyuk Mah.Emek Cad.

Narcity Konutları E1 Blok D:4

Maltepe, Istanbul, Turkey

E-mail: ozlen74@yahoo.com

Key words:

ectropion, eyelid, ichthyosis, surgery

Abstract

Background: Surgical correction of bilateral cicatricial upper and lower eyelid ectropion in an ichthyosis patient remains a challenge in clinical practice.

Main observations: A 24-year-old female patient presented to our clinic with bilateral upper and lower eyelid ectropion. Her skin over her entire body and face was dry and scaly. The diagnosis was cicatricial ectropion related in a patient with ichthyosis. The upper eyelids were treated by retroauricular full thickness skin grafts and upper eyelid lateral tarsal strip procedure. And lower eyelids were treated by cheek transposition grafts and lower eyelid lateral tarsal strip procedure. The upper and lower eyelids were corrected successfully with these surgical procedures.

Conclusions: In patients with ichthyosis skin alterations in the eyelid cause shortening of the anterior lamella, subsequently resulting with ectropion. Successful surgical correction with skin grafts or transposition flaps can be performed to lengthen anterior lamella. Adding lateral tarsal strip procedure to skin grafting helps to maintain a better lid margin apposition. (*J Dermatol Case Rep.* 2011; 5(2): 27-29.)

Introduction

Ichthyoses are a heterogeneous group of inherited disorder characterized by thickening, fissuring and scaling of the skin. It occurs as a result of abnormal keratinization or cornification.^{1,2,3} There are different forms of ichthyosis; ichthyosis vulgaris (autosomal dominant), x-linked ichthyosis, lamellar ichthyosis (autosomal recessive), and congenital ichthyosiform erythroderm.⁴ Lamellar ichthyosis is a rare autosomal recessive disorder affecting 1 in 300.000 people. The most common eyelid abnormality reported is cicatricial ectropion, which is assumed to lead to corneal exposure and ulceration.^{1,5}

In most of the patients surgical treatment is required to relieve symptoms.

Conservative treatment including steroid ointments, methyl cellulose eye drops and humidified environment has failed to achieve a significant and lasting relief.^{4,5} An ichthyosis patient with cicatricial ectropion of both upper and

lower eyelids was treated successfully by retroauricular full thickness autologous skin grafts and cheek transposition flaps, respectively.

Case Report

A 24-year-old female presented with progressive upper and lower eyelid ectropion for the last few years. On physical examination, the skin of her entire body and face was dry and scaly. Lamellar ichthyosis was diagnosed in this patient.

Due to severe keratinization of the skin of the eyelids the anterior lamella was shortened in both of the upper and lower lids leading to cicatricial ectropion. Although these abnormal eyelids both corneas remained clear (Fig. 1A).

Under general anesthesia, both upper eyelid and on another operation day lower eyelids were operated.



Figure 1

Upper and lower lids cicatricial ectropion in the ichthyosis patient before surgery (A), directly after lower eyelid reconstruction (B) and postoperative view 6 months after surgery (C,D).

Upper Eyelid Reconstruction:

Skin crease incision 10 mm from lash line was made. The skin was released to form approximately 3x2 cm skin defect until the lid margin turned to normal position. A full-thickness postauricular skin graft was taken and excess fat was removed from the graft. Haemostasis was secured and the skin defect was closed with interrupted 4/0 silk sutures. The graft was sutured to the defect on the upper eyelid with interrupted 6/0 vicryl sutures.

Additional upper lateral tarsal strip procedure was performed to maintain exact apposition of the upper eyelid margin. Lateral canthotomy was performed, and upper limb of the lateral canthal tendon was identified and cut. A new lateral canthus was formed from the lateral part of the tarsal plate by splitting the upper eyelid by the gray line into anterior and posterior lamella. After shortening the tarsal strip to a appropriate size it was sutured to the periosteum of the lateral orbital rim. The incision was closed with 6/0 vicryl sutures.

A tie over dressing with framycetin-containing wound gauze was maintained 7 days following surgery. Lid skin sutures were removed 7 days after surgery while postauricular skin sutures were removed 10 days after surgery.

Lower Eyelid Reconstruction:

A subciliary incision was made, and lower eyelid skin was released to form a 3x2 cm skin defect. A myocutaneous cheek flap was harvested from the lateral cheek area vertically, and was transpositioned horizontally and sutured to the defect with interrupted 6/0 vicryl sutures.

The vertical defect was closed primarily with interrupted 6/0 vicryl sutures (Fig. 1B).

As the lower eyelids margins were lax this procedure was also combined with a lateral tarsal strip procedure.

A tie over dressing with framycetin-containing wound gauze was maintained 7 days following surgery. Sutures were removed 7 days after surgery.

Upper and lower lid ectropions were corrected successfully with these procedures.

After a 6 months followed-up cosmetic results were found quite good (Fig. 1C,D).

Discussion

Congenital ichthyosis is a generalized hyperkeratinization of the skin. Lamellar ichthyosis is a severe form that involves the entire body, and ectropion is mostly associated with lamellar ichthyosis. Skin alterations in the eyelid cause shortening of the anterior lamella, subsequently resulting in ectropion. This affects the upper eyelid more than the lower and can lead to complications such as chronic palpebral or bulbar conjunctivitis and keratinization or exposure keratopathy.⁶

In ichthyosis the upper eyelid movements are never normal. There is always a certain degree of cicatricial lagophthalmos.¹ For the patients, the presence of ectropion of both eyelids increases the area of palpebral fissure and the rate of tear film evaporation. Also, the absence of the Bell's phenomenon is an additional risk for corneal scarring. In our patient, although ectropion involved both upper and lower eyelids, the Bell's phenomenon was good. It is probably the only explanation for her clear corneas.

Surgical treatment is indicated in most of the patients for ectropion. It is always necessary to lengthen anterior lamella with free skin grafts.^{1,4,5,7} Because of the widespread involvement of the skin and the difficulty in finding disease-free area for transplantation, managing with ectropion in these patients is quite hard. Commonly used sites are the arm, groin, postauricular regions and alternatively prepucce.^{4,5} Extraordinary approaches using mucous membrane graft or maternal skin allograft for correcting cicatricial ectropion has also been reported.^{8,9}

In our case, the postauricular skin grafts were harvested for upper eyelid reconstruction. After suturing the graft to the defect an additional upper eyelid lateral tarsal strip procedure was performed to maintain exact apposition of the upper eyelid margin. The lower eyelids were reconstructed successfully with lateral cheek transposition flaps and lateral tarsal strip procedures. We believe that additional lateral tarsal strip procedure served better lid apposition, because in these longstanding cicatricial ectropions the lid margins seemed to look loosened after the anterior lamellas were corrected with skin grafts and flaps.

In our case a tie over dressing with framycetin-containing wound gauze was maintained 7 days following surgery in both upper and lower eyelids. This helped in getting a viable and moistured graft and flap.

In the short term follow up (6 months) good cosmetic and functional results were achieved. But longer follow up results were not possible to obtain because of patient inconsistency.

Conclusion

In this case presented, adding lateral tarsal strip procedures to the reconstruction of the upper and lower eyelids with skin flaps and grafts helped to maintain a good lid margin apposition.

References

1. Cruz AA, Menezes FA, Chaves R, Coelho RP, Velasco EF, Kikuta H. Eyelid abnormalities in lamellar ichthyoses. *Ophthalmology*. 2000; 107(10): 1895-1898. PMID: 11013195.
2. Turgut B, Aydemir O, Kaya M, Türkçüoğlu P, Demir T, Celiker U. Spontaneous corneal perforation in a patient with lamellar ichthyosis and dry eye. *Clin Ophthalmol*. 2009; 3: 611-613. PMID: 19997563.
3. Chaurasia S, Das S, Ramamurthy B. Microbial keratitis in a case of lamellar ichthyosis. *Int Ophthalmol*. 2008; 28(5): 367-368. PMID: 17828596.
4. Uthoff D, Gorney M, Teichmann C. Cicatricial ectropion in ichthyosis: a novel approach to treatment. *Ophthalm Plast Reconstr Surg*. 1994; 10: 92-95. PMID: 8086369.
5. Hoşal BM, Abbasoğlu OE, Gürsel E. Surgical treatment of cicatricial ectropion in lamellar ichthyosis. *Orbit*. 1999; 19(1): 37-40. PMID: 12045963.
6. Menke TB, Moschner S, Joachimmeyer E, Ahrens P, Geerling G. Congenital ectropion in ichthyosis congenita mitis and gravis. *Ophthalmologie*. 2006; 103(5): 410-415. PMID: 16328488.
7. Singh AJ, Atkinson PL. Ocular manifestations of congenital lamellar ichthyosis. *Eur J Ophthalmol*. 2005; 15(1): 118-122. PMID: 15751249.
8. Nayak S, Rath S, Kar BR. Mucous Membrane Graft for Cicatricial Ectropion in Lamellar Ichthyosis: An Approach Revisited. *Ophthalm Plast Reconstr Surg*. 2011 Feb 22. [Epub ahead of print]. PMID: 21346670.
9. Das S, Honavar SG, Dhepe N, Naik MN. Maternal skin allograft for cicatricial ectropion in congenital ichthyosis. *Ophthalm Plast Reconstr Surg*. 2010; 26(1): 42-43. PMID: 20090485.