

Successful Correction of Ichthyosis-related Ectropion by Autografts

Running Title: Surgical Repair of Ectropion in Ichthyosis

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Abstract

Ectropion is a rather frequent complication of ichthyosis with negative functional and esthetic impact. Lid surgery can provide significant improvement. We report on a series of four ichthyosis patients with ongoing bilateral eye problems despite intense medical treatment (mean age 27.8 ± 14.1 years). All patients suffered from lagophthalmos. Two of the patients had only lower lid ectropion. In two of the patients' ectropion was forming on the upper lid in addition to the lower lid when closing the eye. In three of four patients, ectropion was repaired by skin grafts from the supraclavicular region. In a child with lower and upper lid ectropion prepuce was used for repair surgery. At the end of the follow-up period of about 23.0 ± 12.7 months, none of the patients revealed lagophthalmos or corneal exposure. No intraoperative or postoperative complications have been observed in our patients. Ichthyosis patients with ectropion resistant to medical treatment, benefit from surgical treatment with full layer autografts. In boys, prepuce can be successfully used as autograft.

Keywords: ectropion, eyelid, ichthyosis, skin graft, surgery, treatment.

Introduction

Ichthyosis is a hereditary and heterogeneous group of hereditary disorders of cornification which are characterized by hyperkeratosis and a fish scale look.¹ Ichthyosis is divided into sub-groups such as ichthyosis vulgaris, x-linked ichthyosis, Harlequin ichthyosis, lamellar ichthyosis and congenital

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ichthyosiform erythroderma based on hereditary characteristics, clinics and histopathological appearance.² The most widespread ocular manifestation is cicatricial ectropion and mostly seen as lamellar type.^{3,4}

Cicatricial ectropion can be congenital or can be seen in the further phases of life.⁵ As characteristics of ichthyosis disease, trans epidermal dehydration as well as contractions and loss of elasticity in eyelid skin arise as a result of compensatory hyperproliferation.^{2,6} Tissue loss and vertical shortening are seen in anterior lamella and thus ectropion arises.⁵⁻⁷

Other ocular manifestations such as lagophthalmos, conjunctival hyperemia, exposure keratopathy, recurrent corneal ulcer mostly develop due to cicatricial ectropion.^{3-6,8} Furthermore, the disease itself may also cause ocular manifestations with systemic involvement.³ Patients complain about photophobia, pain, tearing, foreign body sensation and loss of vision.⁷

Corneal damage arising from ectropion can be eased with moisturizers and synthetic tears while chronic inflammation can be eased medically with steroids and immuno-modulators. However, surgery is needed to cure advanced ectropion.^{3,4,6,9,10} Full-thickness skin defects are required in surgical treatment, but it is hard to find healthy tissues since all skin is affected by the disease. Inner arm, retro auricular and supraclavicular region,³ penile skin (prepuce),⁸ groin^{3,11} and mucous membrane grafts⁷ are successfully used in this sense.

In this article, we have aimed to present four cases we have treated the cicatricial ectropion by using autografts prepared from supraclavicular region and prepuce.

Materials and Methods

This study protocol was approved by the local Institutional Review Board and was conducted in accordance with the Declaration of Helsinki. Informed consent was obtained from all individual participants included in the study.

This study includes 8 eyes with an ectropion of four ichthyosis patients. The age range was 7 to 38 years with a mean age of 27.8 ± 14.1 years. All patients had complaints such as foreign body sensation, watering of eyes and eye burning. All suffered from lagophthalmos, conjunctival hyperemia and corneal punctate epitheliopathy intensifying especially in the inferior region. One patient (Patient 3) had lateral pterygium and intense lacrimation. In all patients' posterior segment examination and intraocular pressure were normal limits and corrected visual acuity was 20/20 with the Snellen chart. Clinical characteristics of the patients are listed in Table 1.

Two of the patients had only lower lid ectropion. In two of the patients, ectropion was forming on the upper lid in addition to the lower lid ectropion when closing the eye.

An adult patient with lower and upper lid ectropion (Patient 1) underwent surgery in general anesthesia. Lower and upper lid ectropion's were repaired using skin grafts extracted from the supraclavicular region (Figure 1). Two patients with only lower lid ectropion (Patient 2 and 3) were treated in local anesthesia using skin grafts extracted from the supraclavicular region (Figure 2 and 3).

A child with lower and upper lid ectropion (Patient 4) underwent combined upper and lower lid ectropion repair using prepuce extracted under general anesthesia (Figure 4). All of the patients had lagophthalmos

During surgical treatment of lower lid; the lower lid was applied sub-ciliary incision. Lid skin was separated from orbicularis muscle and surrounding tissues and lid was released. The full-thickness skin graft was sutured to this area one by one by using a 6-0 prolene suture. Two small incisions were made on the graft so that hematoma which can accumulate under the graft and leakage were drained. Two

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or three reciprocal sutures placed on the graft were left long and they were tied on the sponge to apply pressure on the graft. In upper lid surgery, the incision was made on the current lid crease of the patient or if the upper lid crease couldn't be detected, at a distance of 8 mm from the upper lid edge. Lid skin was separated from orbicularis muscle and surrounding tissues and the lid was released. The skin graft was sutured by 6-0 prolene suture. The graft was sutured with the same method applied on lower lid skin graft. Extracted skin grafts were prepared 1/3 larger than the lid defects created by releasing the lid. Sutures were removed about one week after surgery based on the recovery of the tissue.

Statistical analyses were performed using the SPSS software version 16 (SPSS Inc., Chicago, IL, USA). Descriptive statistical methods (mean, standard deviation, median, frequency, ratio, minimum, maximum) were used evaluating of the data.

Results

All patients have been followed up for a minimum of 6 months. The mean follow-up period was 23.0 $\pm$ 12.7 months. After the operation, improvement in eye signs and symptoms was observed in all patients (Figures 1 to 4). There was no patient who showed lagophthalmos during the follow-up period postoperatively. No complications have been observed on the eyelids or donor site during or after the operation.

Discussion

Ichthyosis is a spectrum of skin diseases frequently related to cicatricial ectropion and lagophthalmos as ocular involvement.^{3,4} Ectropion is most frequently observed in lamellar ichthyosis and seen in 45%

to 80% of the cases.^{3,5,12} It causes skin rashes and dryness, heavy contractions, and lagophthalmos by shortening anterior lamella.⁵ Independent of exposure, the disease itself can lead to ocular manifestations as well.³ As a consequence of exposure conjunctival hyperemia, keratopathy, and recurrent corneal ulcer may develop.^{3,4,6,8} Cinar et al. reported spontaneous bilateral corneal perforation in an 8-month-old baby with lamellar ichthyosis. Defect closure was realized with a two-layered amniotic membrane.¹³ Corneal damage is related to upper lid ectropion.^{3,6} In moderate cases, conservative approaches such as moisturizers and artificial tears are recommended while in advanced damages surgical treatment is required.^{3-6,8-10,14}

When surgery is necessary it should be performed early to avoid loss of vision and possible complications.³ The aim of surgery is to lengthen the anterior lamella and enable the eyelids to close.¹² However, in ichthyosis patients it is hard to find intact donor skin to be used as a graft. In these patients possible donor sites are antecubital fossa, supraclavicular, area postauricular region, groin, and foreskin.^{5,8}

Ozgul et al. used retro-auricular full-thickness skin graft to repair the upper eyelid of an ichthyosis patient with ectropion on the bilateral upper and lower eyelid and they used transposition grafts from the cheek to repair lower lids.⁶ Das et al. applied a maternal postauricular allograft on a 6-month-old patient with involvement of the whole body. At the end of the 10-month follow-up period, they observed that the graft had completely been taken without immunosuppression. The ectropion was successfully repaired.¹⁴

In male patients, penile foreskin is preferred since it does not contain any hair follicles and is loosely attached to the penis.^{5,8} We too used prepuce successfully to repair ectropion of lower and upper eyelids in a 7-year-old boy.

Engineered human skin equivalent (Appligraf) is of limited value.¹⁵ Mucous membrane grafts extracted from the cheek can be used to repair lower eyelid ectropion.^{7,16} Alkatan et al. applied mucous membrane graft to a 21-year-old patient with ectropion in lower eyelids. Although metaplasia and keratosis have been observed in the graft, dysplasia and malignancy did not occur. They reported a good recovery and cosmetic look.⁷ Supraclavicular skin grafts can be used to lengthen the anterior lamella in cicatricial diseases with ocular involvement.^{17,18} Bayhan et al. applied the autograft they extracted from the supraclavicular region to both upper and lower eyelids of a 6-year-old girl with ichthyosis-related ectropion.¹⁹ We have successfully applied supraclavicular skin grafts to lower eyelids of 2 male adults, and to lower and upper eyelids of a male adult. A concern is possible tissue contraction, therefore a graft 1/3 larger than the real defect is recommended.³

Attention has to be paid to graft extraction and suturing. Ozgul et al. applied a lateral tarsal strip procedure after repositioning the graft. Otherwise, a lengthened anterior lamella can cause a loose look of the edges of the eyelids after surgery.⁶ To correct horizontal eye lid laxity lateral tarsal stripe should be included.⁸

In conclusion, in cases in whom conservative treatment is not sufficient, autograft surgery is recommended early to repair ectropion and prevent complications. In boys foreskin autograft can be successfully applied after informed consent of the parents. Our technique offers an excellent functional and esthetic outcome and prevents stigmatization.

Table 1: Clinical data of the patients

	Patient 1	Patient 2	Patient 3	Patient 4
Type of Ichthyosis	Ichthyosis vulgaris	Ichthyosis vulgaris	Ichthyosis vulgaris	Lamellar ichthyosis
Age (years)	32	38	34	7
Follow-up (months)	28	6	22	36
Sex	M	M	M	M
Ocular conditions	Bulbar and palpebral conjunctival hyperemia, corneal punctate epitheliopathy	Conjunctival hyperemia, Corneal punctate epitheliopathy	Conjunctival hyperemia, punctate keratopathy in inferior cornea, pterygium, lacrimation	Conjunctival hyperemia, corneal punctate epitheliopathy
Eyelid side	Right and left eye lower and upper lids	Right and left eye lower lids	Right and left eye lower lids	Right and left eye lower and upper lids
Graft (donor) side	Supraclavicular	Supraclavicular	Supraclavicular	Prepuce
Complication	None	None	None	None
Anesthesia	General	Local	Local	General

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Figure Legends

Figure 1. Patient 1: Ectropion of lower and upper eyelids. (a) Eyes open. (b) Eyes closed. (c) Four days after surgical treatment of upper and lower eyelids ectropion. (d) Result 15 months after surgery.

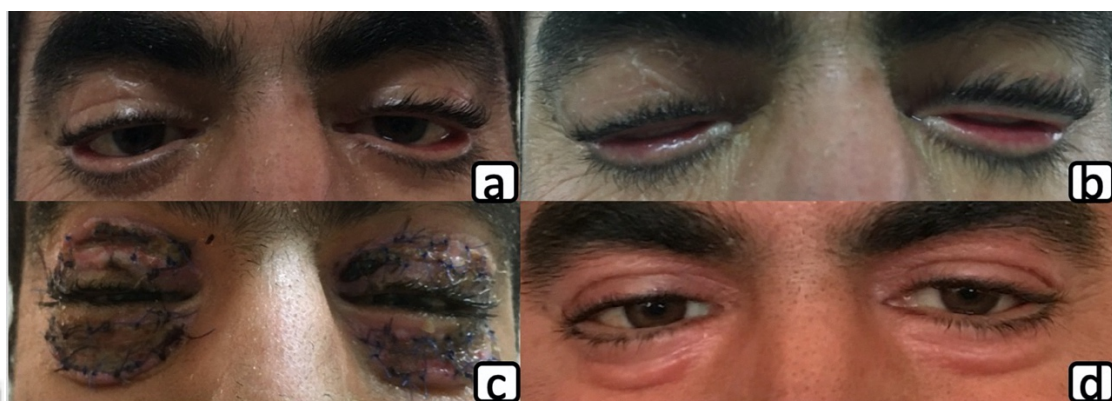


Figure 2. Patient 2: (a) Lower lids with cicatricial ectropion before surgery. (b) Five days after surgery.



Figure 3. Patient 3: (a) Preoperative appearance of the bilateral lower eyelids cicatricial ectropion. (b) Three days after surgical correction. (c) Ten months after correction with supraclavicular graft.



Figure 4. Patient 4: A child presenting with ichthyosis and cicatricial ectropion of both upper and lower lids. (a) Eyes open. (b) Eyes closed. (c) First day after surgery showing full-thickness foreskin graft. (d) Fourteen months postoperatively with excellent functional and esthetic outcome.

