

Progressive Ectropion due to Lamellar Ichthyosis: A Therapeutic Proposal

Marcus Vinícius Martins Collares, MD¹

Paula Mancopes²

Márcio Hoffman Rigo, MD³

Roberto Corrêa Chem, MD⁴

Rinaldo De Angeli Pinto, MD⁵

- 1] Maxilofacial Plastic Surgeon of Hospital de Clínicas de Porto Alegre's Plastic Surgery Service, Surgery Professor in the PGC at the FAMED – UFRGS.
- 2] Student of Medicine at the Universidade Federal do Rio Grande do Sul.
- 3] Plastic Surgeon, former resident of Hospital de Clínicas de Porto Alegre's Plastic Surgery Service.
- 4] Plastic Surgeon, Professor of Medicine at the Universidade Federal do Rio Grande do Sul.
- 5] Plastic Surgeon, Professor of Medicine at the Universidade Federal do Rio Grande do Sul. Head of Hospital de Clínicas de Porto Alegre's Plastic Surgery Service.

Address for correspondence:

Marcus Vinícius Martins Collares, MD

R. Hilário Ribeiro 202/406
90510-040 – Porto Alegre – RS
Brazil

Keywords: Ichthyosis; cicatricial ectropion; skin graf.

ABSTRACT

A 48-year-old male patient suffering congenital ichthyosis, bilateral ectropion of the lower eyelids, erythroderma, and diffuse desquamation. The congenital lamellar ichthyosis is associated with ectropion and exposure keratitis. Concha-cartilage, oral mucous membrane, and prepuce skin grafts were successfully used for eyelid repositioning and eyeball protection.

INTRODUCTION

Congenital lamellar ichthyosis (CLI) is a recessive autosomal disease characterized by mild erythroderma and extensive, fine, dark and flat scales that covers all body surface. The CLI is frequently associated with severe ectropion. The ectropion exposes the cornea and this may cause corneal abrasion due to keratinized squamous epithelium friction. Repetitive chronic aggression to the cornea may cause blindness, thus eyelid reconstruction is necessary^(1, 3).

Aesthetics and function should be considered in reconstruction surgeries. Reconstruction of the eyelid requires skin, mucous membrane and a supporting structure. Most of the available reconstruction techniques were described for tumor resections. These techniques include: cutaneous flaps, labial and oral mucous membrane grafts, mucoperiosteal graft from the palate, irradiated tarsal graft, aorta graft, contralateral eyelid composite graft, and nasal and auricular cartilage graft^(4, 5).

Eyelid reconstruction in CLI patients involves an additional difficulty: the lack of feasible skin throughout the body. There are few data regarding ectropion repair in CLI patients in the medical literature.

The purpose of this paper is to report an eyelid reconstruction in a CLI patient with bilateral ectropion and to evaluate the results.

CASE REPORT

A 48-year-old male patient, with CLI could not close his eyes and had presented recurrent ulcers on the cornea in the last 4 years. They were caused by an ectropion in the left lower eyelid and a retraction in the right lower eyelid (Fig. 1). He also presented an erythroderma and diffuse desquamation. In January, 1994 he had a corneal perforation of the left-eye that led to blindness. He was referred to the Plastic Surgery Department to treat the eyelid defect in the right eye in order to preserve the right eye function.

A right eyelid reconstructive surgery was performed in September 1994. The surgical technique consisted of subciliary incision, skin undermining to enable cranial sliding of the lower eyelid (Fig. 2), release of orbicular muscle insertion at the orbital brim, lifting of the orbicular muscular flap (Fig. 3), placement of conchal-cartilage graft under the muscular flap to support the eyelid, and an oral mucous membrane graft to cover the skin gap over the eyelid muscle. Seven days after the surgery, a necrosis of the oral mucous membrane graft was observed. A surgical débridement was performed, and four days later a new oral mucus membrane graft was placed over the granulation tissue. Eighteen days after surgery, the surgical wound was healing well, and the patient could almost completely close his eyes.

Ectropion of the left eye was repaired one year later. The same technique described above was used. An incision on the left lower eyelid was made, the orbicular muscle was exposed, and a cartilage from the left external ear was obtained. The cartilage was sutured under the orbicular muscle and covered with a skin graft from the prepuce. In November 1995, the patient could completely close his left eye (Fig. 4), and four years later he could close both eyes.

DISCUSSION

Congenital lamellar ichthyosis is also known as type-4 keratinization disorder. It is a rare recessive autosomal

disease characterized by diffuse desquamation of the skin. The scales are large and dark brownish. There is also an erythroderma and a severe ectropion. The histology reveals hyperkeratosis and hypergranulosis. The cellular turnover is practically normal and the prognosis does not change as the patient ages⁽⁶⁾.

The excessive skin dryness causes eyelid retraction and ectropion in CLI patients. Ectropion and lagophthalmos produce ocular disturbances due to cornea exposure. As shown by several studies, the damaged cornea can heal and become vascularized or can perforate, thus it is important to develop adequate surgical techniques to treat the ectropion⁽⁷⁾.

In 1973, Schindle and Leone⁽⁷⁾ used a retroauricular skin graft to repair a right eye ectropion in an 8-year-old CLI patient, and a left retroauricular skin graft to correct the left eye ectropion. It was not necessary to use cartilage to support the eyelids. Six months after the surgery, the patient presented a visual acuity of 20/20, no corneal disturbances, and no need for medication to the eyes.

In 1994, Uthoff *et al*⁽⁸⁾ used prepuce skin to repair the four eyelids in a 15-year-old CLI patient with bilateral ectropion. The prepuce skin was the only source of unaffected skin in that patient. The sleeve technique was applied and the prepuce skin was divided into four triangles to be used as grafts after the ectropion release. A partial and temporary tarsorrhaphy suture was used to enable synechiae formation between upper and lower eyelids. This allowed the patient to see while taking off the graft, and prevented the ectropion recurrence. The synechiae was undone 3 months after the surgery.

We reported a case of long-term evolution CLI with severe ectropion and ocular complications. A technique that applies a cartilage to support the lower eyelids was required. The prepuce skin was the only source of unaffected skin, but the oral mucous membrane was an effective skin substitute.

The long-term evolution aggravated the ectropion, which required a technique more complex than a simple skin graft technique.

CONCLUSION

The surgical technique we employed is effective in treating ectropion. A normal palpebral closure and an ocular symptoms improvement were observed in a 4-year follow-up.

REFERENCES

1. SAMPAIO SAP *et al.* Afecções Congênicas e Hereditárias. In: *Dermatologia Básica*. 3. ed. Artes Médicas, 1989. p. 451-73.
2. WILLIAMS ML, ELIAS PM. Heterogeneity in autosomal recessive ichthyosis. *Arch. Dermatol.* 1985; 121: 477-88.
3. WILLIAMS ML, ELIAS PM. Elevated nalkanes in congenital ichthyosiform erythroderma – phenotypic differentiation of two types of autosomal recessive ichthyosis. *J. Clin. Invest.* 1984;74: 296-300.
4. PERRY MJ *et al.* Lower eyelid reconstruction using pedicled skin flap and palatal mucoperiosteum. *Dermatol. Surg.* 1997; 23: 395-98.
5. JORDAN DR, ANDERSON RL. Eyelid reconstruction with irradiated human tarsal plate and aorta. *Int. Surg.* 1997; 82: 350-58.
6. LYNCH PJ. Generalized Disorders of Cornification: The Ichthyosis. In: *Principles and practice of dermatology*. 2. ed. Churchill Livingstone, 1996. p. 379-96.
7. SCHINDLE RD, LEONE CR. Cicatricial ectropion associated with lamellar ichthyosis. *Arch. Ophthalmol.* 1973; 89: 62-4.
8. UTHOFF D, GORNEY M, TEICHMANN C. Cicatricial ectropion in ichthyosis: A novel approach to treatment. *Ophthalmic Plast. Reconstr. Surg.* 1994; 10: 92-5.



Fig. 1 – Preoperative aspect. Cicatricial ectropion of the left lower eyelid and right superior eyelid retraction. Sequela from the left eye cornea perforation.

Fig. 1 - Aspecto pré-operatório. Ectrópio cicatricial de pálpebra inferior esquerda e retração palpebral à direita. Sequêla do rompimento da córnea do olho esquerdo.



Fig. 2 – Subciliary incision and skin undermining to enable upward sliding of right lower eyelid.

Fig. 2 - Incisão subciliar e descolamento da pele para permitir o deslizamento cranial da pálpebra inferior à direita.



Fig. 3 – Release of orbicular muscle insertion and lifting of orbicular muscle flap.

Fig. 3 - Liberação da inserção do músculo orbicular no rebordo orbitário e elevação do retalho do músculo orbicular.



Fig. 4 –Palpebral occlusion 14 months after the first surgery.

Fig. 4 - Fechamento da fenda palpebral visto no exame 14 meses após a primeira cirurgia.