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Oculoplastic Approach to Congenital Cutis Laxa Syndrome

Ahmet Tas · Fatih C. Gundogan · Tolga Kocaturk · Salih Altun · Yalçın Bayram

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Abstract Cutis laxa is a rare congenital or acquired disorder of elastic tissue, characterized by loose skin with folds and multiple internal organ involvement, which may cause life-threatening complications. We present a patient with cutis laxa syndrome who had cross eyelids with esotropia. Bilateral lateral canthal tendon repositioning and bilateral medial rectus recession procedures were performed in a single session. The patient had acceptable eyelid and globe cosmesis after the procedure.

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Keywords Cutis laxa syndrome · Epiphora · Strabismus

A. Tas (✉) · T. Kocaturk
Department of Ophthalmology, Ankara Mevki Military Hospital, Altındağ, Ankara, Turkey
e-mail: anzhtas@hotmail.com

F. C. Gundogan
Department of Ophthalmology, Etimesgut Military Hospital, Ankara, Turkey

S. Altun
Department of Ophthalmology, Kayseri Military Hospital, Kayseri, Turkey

Y. Bayram
Department of Plastic and Reconstructive Surgery, Erzurum Military Hospital, Erzurum, Turkey

Introduction

Cutis laxa syndrome, which was described by Alibert for the first time in the literature in 1832 [1], is a heterogeneous group of connective tissue disorders, characterized by loose skin and variable systemic involvement. The fundamental abnormality is the elastic tissue disorder that involves the skin and other systems [2]. Congenital cutis laxa is inherited as an X-linked, autosomal recessive or dominant trait [3, 4]. and often occurs as part of a syndrome because elastin is important for the structural integrity of other tissues, including bone, ligaments, cartilage, and the walls of large blood vessels [3, 5]. Acquired cutis laxa can occur in association with drug ingestion, solid and hematogenous neoplasia, and inflammatory skin disorders [6–8]. The clinical features are variable in both the congenital and acquired forms, and they range from localized to severe and potentially life-threatening systemic disease with genitourinary (vesicourinary tract diverticula), gastrointestinal (diverticula), cardiovascular (aortic aneurysms), skeletal (hypermobile joints), and respiratory involvement (pulmonary emphysema, bronchiectasis, pulmonary artery disease, severe congenital lung disease) [9–14].

The characteristic clinical pattern is lax and pendulous skin with redundant folds resulting from loss of elastic tissue in the dermis. Facial features include an aged appearance, long philtrum, high forehead, large earlobes, and beaked nose [9]. Skin laxity can be generalized or localized, with or without systemic involvement [5]. Although there are some reports about the surgical correction of cutis laxa, none of them presented a concomitant surgical procedure performed with primary surgery of the skin redundancy.

The aim of this study is to present the concomitant surgical treatment of a patient with cutis laxa and strabismus as well as discuss the surgical outcome.

Case Report

A 12-year-old girl was admitted to the Ophthalmology Department of Ankara Mevki Military Hospital with the chief complaints of cross eyelids, watery eyes, and alternating esotropia (Fig. 1a, b). She was the third child of a healthy distant relative couple. The family history was negative for such skin and/or eye problems. The patient did not report any chronic topical or systemic medication. There was no previous history of trauma, surgery, or eyelid edema. There was a marked excess of upper-eyelid skin (Fig. 1c) with inferior displacement of lateral canthi and scleral show (Fig. 1b). The patient had alternating esotropia with bilateral inferior oblique muscle overaction. The lateral canthus was 4 mm lower than the medial canthus which caused epiphora. The temporal palpebral fissure apertura was higher than the nasal aperture bilaterally due to the palpebral skin laxity and inferior displacement of the lateral canthi (Fig. 1a, b). Best corrected visual acuities were 20/40 with $+1.00 \times 170$ D refraction in the right eye and 20/32 with -1.50×90 refraction in the left eye. Intraocular pressures (IOP) were 12 mmHg in both eyes. Biomicroscopic examination showed a mild injection of the temporal and inferior bulbar conjunctiva. Anterior segments and dilated fundus examination were unremarkable.

Physical examination revealed hypermobile joints (Fig. 2a) and excessively loose redundant skin, especially in the neck (Fig. 2b) and in the abdominal area (Fig. 2c), which are the typical appearances of cutis laxa.

The patient was operated on under general anesthesia. Bilateral inferior oblique myectomy and 6.0 mm of bimedial rectus muscle recessions were performed to correct esotropia. A Tripier flap was planned to correct the inferior displacement of the lateral canthus. A Tripier flap is a myocutaneous transposition flap that utilizes redundant tissue on the upper eyelid to repair defects of the lateral inferior eyelid. After the first description by Tripier in 1889, several variations of the procedure were performed [15–17]. In our case, a laterally based upper eyelid flap was designated and marked with gentian violet on the lateral upper eyelid. The incision of the flap was deepened through the orbicularis muscle (Fig. 3a). Then the lateral orbital rim periosteum was exposed at the level of the lateral canthal tendon. A 5/0 prolene suture was passed through from the lateral canthal tendon and the tendon was cut away from its orbital insertion. Then, the lateral one fourth of the inferior orbital septum was detached from the inferior orbital rim to be able to transfer the lateral canthal tendon to a higher position. Following the dissection, lateral canthopexy was performed and the lateral canthal tendon was sutured to the lateral orbital rim periosteum 5 mm higher than its original insertion. The Tripier flap was transferred from the upper lid to the lower-lid defect. Skin and subcutaneous tissue were closed with interrupted 6/0 Vicryl sutures (Fig. 3b). Postoperative recovery was uneventful with normal healing. The appearance of the patient's upper- and lower-eyelids was acceptable and the esotropia was successfully corrected at 1 year postoperatively. The lateral canthus was at a normal position and the scleral show was decreased to an acceptable level (Fig. 4).

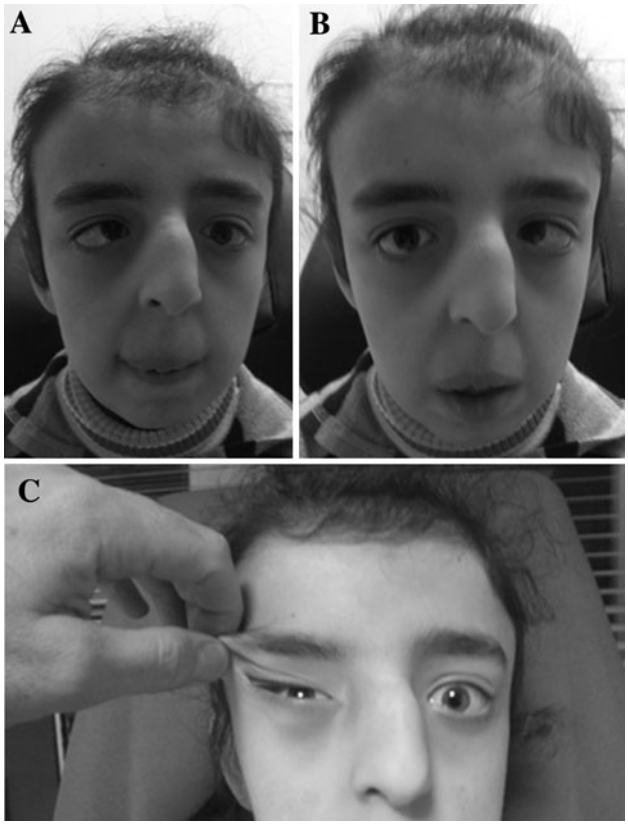

Fig. 1 Cross eyelids, alternating esotropia, beaked nose, and excess upper-eyelid skin

Discussion

Only a few articles in the literature have focused on the role of plastic surgery to solve the cosmetic problem in cutis laxa syndrome. To the best of our knowledge, this is the first case of congenital cutis laxa syndrome with cross eyelids and strabismus corrected in a single operation. In this report, we aimed to present successful combined strabismus surgery and lateral canthal tendon repositioning with a Tripier flap procedure. These procedures corrected the position of the globe and eyelids, and they helped to construct a cosmetically acceptable eyelid appearance.

It is important to differentiate cutis laxa syndrome from other diseases with hyperextensible skin, such as Ehlers-Danlos syndrome and pseudoxanthoma elasticum, which is notorious for its poor wound healing and unacceptable scar

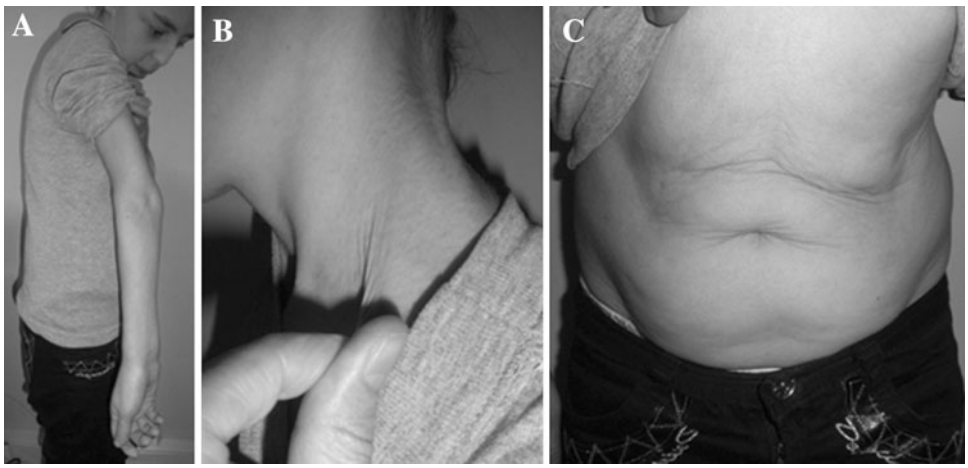

Fig. 2 Typical features of cutis laxa can be seen as a hyperextensible elbow (**a**), loose skin at the neck (**b**) and abdominal area (**c**)

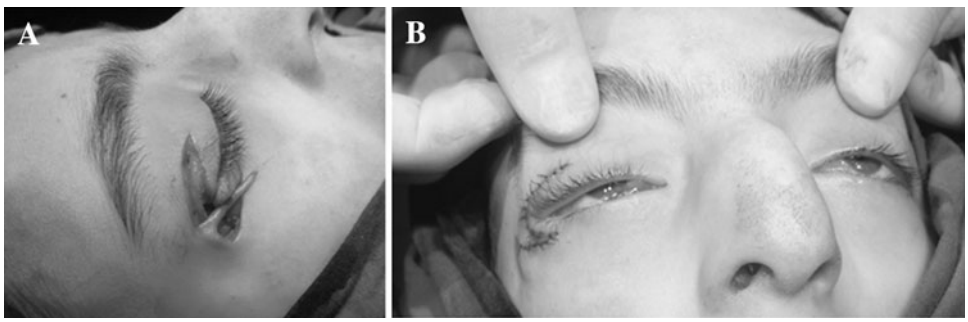

Fig. 3 **a**, **b** Tripier flap was used to correct the excess upper-eyelid skin, support the lower eyelid, and suspend the lateral canthus

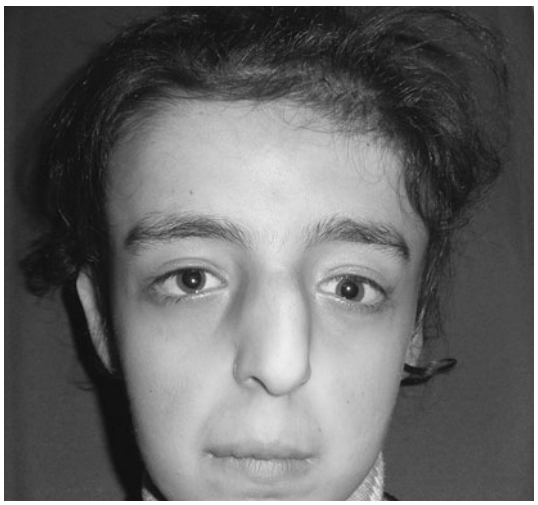

Fig. 4 The postoperative appearance of the eyelids and globe at the first year's examination

formation [3, 5, 18, 19]. A skin biopsy can be helpful in differentiating these conditions. Patients with Ehlers-Danlos syndrome and pseudoxanthoma elasticum may present with vascular abnormalities [20]. In contrast to those diseases, patients with cutis laxa syndrome are good candidates for surgical procedures and excellent results may be obtained. However, realistic expectations in most circumstances are necessary, and it should be kept in mind that future revisions may be necessary because of the progressive nature of the disease. The patient presented here was diagnosed with cutis laxa and we considered her to be a good candidate for the surgical procedure. The main complaints of our patient were epiphora, inferior lateral canthal displacement, and strabismus. Thus, surgery was planned to correct all these deformities in a single procedure. During the surgery of the soft tissues, extreme caution was used to reduce the load on the inferior eyelid and to suspend the lateral inferior eyelid to a higher point of the periosteum. In our case the Tripier flap would have caused an increase in the lateral inferior eyelid load and thus exaggerate the lateral inferior scleral show. This was a potential problem. However, intraoperatively, we saw that the suspension effect of the Tripier flap over the lateral inferior eyelid caused it to have a higher position. For this reason, we did not need to perform any other procedure for the scleral show problem.

Both the surgical procedure and the postoperative period were uneventful and the surgical outcome was satisfactory. Preoperatively, we thought that nonsurgical techniques for tissue tightening, such as radiofrequency energy, would not be an ideal procedure for our patient because of the hereditary soft tissue problems that may impair wound healing. However, combined treatment strategies can be used in selected cases [21].

Another important factor in decision-making with these patients is the timing of the surgery. Reports in the literature suggest that early surgery for children with cutis laxa, while school age, reduces their difficulties in social relationships [22]. Although it is advocated by some authors that the patients' growth can fill out the skin envelope, we preferred the operative approach because of functional impairment caused by the deformity [23]. It is reported that with age, the changes in the skin of cutis laxa patients tend to become less severe and the aging appearance of the face stabilizes. Moreover, fibrosis secondary to surgical dissection contributes to stabilization of the skin [19]. Judging from the 12-month follow-up examination, we think that the surgical outcome of our patient will possibly be long-lasting and revision surgery may not be needed.

In conclusion, surgery in cutis laxa may provide important functional and aesthetic benefits to the patient. Periorbital skin laxity and strabismus can be surgically corrected simultaneously. However, the decision for surgery should be made cautiously in terms of timing.

Disclosures The authors have no conflicts of interest to disclose.

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