CASE REPORT

Acquired Localized Cutis Laxa: A Case Report and the Role of Plastic Surgery

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Abstract
Cutis laxa is an uncommon connective tissue disorder affecting the elastin fibers leading to lax and pendulous skin and in generalized form can present with systemic involvement. Congenital cutis laxa is common in comparison to acquired cutis laxa and has varied inheritance patterns. Treatment is chiefly observation in congenital cutis laxa, and there is a paucity of literature on surgical management in acquired cutis laxa. We report a rare case of acquired localized cutis laxa with a review of literature on the role of plastic surgery in this condition.

KEY WORDS: Acquired, cutis laxa, elastin, plastic surgery

Introduction
Cutis laxa is a rare connective tissue disorder affecting skin with decreased elastin in the dermis, leading to a characteristic appearance of pendulous skin with decreased elasticity and resilience of the skin. The disease can manifest as congenital or acquired forms, the congenital variant being more common compared to the latter. The congenital variant can present in autosomal recessive, dominant, or X-linked inheritance. Both the forms can present either as a generalized or as a localized entity.

In the localized form, there is only skin involvement, but with increasing severity, the chances of systemic affliction in the form of gastrointestinal (diverticulae, hernias), genitourinary (diverticulae), cardiovascular (aneurysms), and pulmonary involvement are increased. In its most severe form, cutis laxa is lethal in the early postnatal life secondary to pulmonary complications. Acquired cutis laxa is rare compared to congenital form and has an underlying cause, leading to onset such as urticaria, exposure to drugs (penicillamine, isoniazid, and penicillin), and neoplasms (usually hematological) in rare instances.

Acquired cutis laxa usually starts in the face and progresses downward and is manifested in the form of lax skin with redundant folds. There is a paucity of literature on the surgical procedures for cutis laxa. Acquired cutis laxa localized to face is extremely rare, and there are only six published case reports to our knowledge. We report a case of acquired cutis laxa localized to face and managed by surgery along with a review of literature of this uncommon condition.

Case Report
A 26-year-old male presented to our outpatient department with the complaints of saggy and inelastic skin under his eyes on both cheeks for 4 years. It started as ill-defined, erythematous lesions over the malar region which stabilized and later led to skin changes in the form of wrinkles over both cheeks. On examination, the patient showed aging facies with saggy skin under both eyes which was inelastic and wrinkled. No other body parts were involved. No hypermobile joints were noted. There was no history of urticaria or eruptions, drug intake, respiratory complaints, or history suggestive of vasculitis. No positive family history about similar complaints could be elicited.

On further workup of the patient, hemogram, serum electrophoresis, and erythrocyte sedimentation rate were found within normal range. Chest radiograph and pulmonary function tests revealed no respiratory anomaly. Echocardiography of the patient was normal and there was no involvement of abdominal organs on

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ultrasound. Skin biopsy from the involved segment was taken and subjected to hematoxylin and eosin (H and E) and van Gieson's staining which showed a severe reduction in elastin in all the layers of dermis [Figure 2].

The patient was taken up for surgery under local anesthesia. A crescent-shaped area of the affected skin was marked for excision and infiltrated with local anesthetic with adrenaline [Figure 3]. The area of abnormal skin was excised, leaving a defect with unequal margins over both malar areas. Wide undermining of the cheek skin was done and suturing begun from the medial to the lateral margin of the defect. The wound was closed taking unequal bites from both margins to minimize the inevitable dog-ear at the end of the incision. The dog-ear left at the lateral end of the defect, toward the end of the malar area, was closed by excising the Burrow's triangles formed due to the excess skin of the lower margin, thus resulting in an esthetic closure of the wound. The closure was done in two layers after ensuring adequate hemostasis, and a light compressive dressing was applied at the end of the procedure. Suture removal was done at the end of 6 days and the scar was strapped with Steri-Strips to prevent stretching. The patient was advised appropriately regarding scar management and was followed up at regular intervals. At the end of 6-month follow-up, the patient showed no recurrence and was satisfied with the esthetic outcome of the procedure [Figure 4].

**Discussion**

Acquired cutis laxa is rarer than its congenital counterpart and is more often symptomatic in early adulthood. It has an insidious onset and may be associated with urticaria or drug intake. The onset is usually preceded by inflammatory dermatoses or less commonly a hematological malignancy. Early onset is associated with better prognosis, and the lesions are more likely to fill out with growth.\(^5\)

Majority of the acquired cases come under the generalized category (Type I), which usually begins in adulthood and follows an urticarial reaction, as a hypersensitivity reaction to insect bite, following drug intake or a malignancy. The localized (Type II) acquired cutis laxa, also called Marshall's syndrome, is extremely rare and is usually seen at a younger age and is usually associated with insect bite.\(^6,7\)

Low levels of lysyl oxidase activity and a higher level of cathepsin G along with decreased α1 antitrypsin are predicted to lead to decreased elastin in the dermis, especially the papillary dermis.\(^8\) Missense mutations in the elastin (ELN) and fibulin-5 (FBLN5) genes\(^9\) are associated with an increase in the susceptibility of elastin fibers to inflammatory degradation. Another hypothesis has been put forward, suggesting that elastolysis results from elastase release from neutrophils and macrophages.\(^10\)

Diagnosis is usually by clinical suspicion and is confirmed by skin biopsy and histopathological examination by H and E and van Gieson's staining. Histopathological studies are chiefly aimed at differentiating the lesion
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from other connective tissue disorders such as Ehlers–Danlos syndrome, pseudoxanthoma elasticum, and postinflammatory elastolysis.

Treatment of cutis laxa is largely conservative. In the congenital variant, spontaneous resolution can be anticipated and hence observation is preferred. No pharmacological agent has been known to stop the progression of the disease. Dapsone has been used to control the inflammatory swelling but failed to control the disease. Unlike other connective tissue disorders such as Ehlers–Danlos syndrome, vascular fragility is not encountered in cutis laxa and hence surgery is not contraindicated as it does not affect wound healing.

There are very few reports on surgical management of acquired cutis laxa in the literature. Tas et al. reported their experience in a child with cutis laxa by oculoplasty with Tripier flap and had a satisfactory postoperative result at the end of 1 year. They also suggested the differentiation of the condition from other connective tissue disorders such as Ehlers–Danlos syndrome and pseudoxanthoma elasticum by a skin biopsy as they are associated with a poor outcome following surgical intervention.

Riveros et al. published their experience with a case of localized acquired cutis laxa in a young woman in her late 20s without prior inciting cause. They reported satisfactory outcome in her following correction of the earlobes, rhytidectomy, and bilateral blepharoplasty. They also stated that plastic surgical correction often is the only resort, with rhytidectomy being the first procedure of choice.

In another article by Mitra et al., the authors gave description of two patients with acquired cutis laxa and also reported satisfactory outcome with a fine scar following reconstructive surgery in one of the patients and also cautioned about the need for revision surgeries in future.

Nahas et al. in their study on the role of plastic surgery in congenital cutis laxa described a patient in her 20s who underwent two rhytidectomies, the first rhytidectomy done along with a superficial muscular aponeurotic system procedure, with a poor result after the first procedure and a satisfactory one after the second time. The patient was lost to follow-up and reported 10 years later with signs of recurrence. They opined that plastic surgery had a definite role in allaying the social and psychological issues associated with the skin laxity and also brought the patients’ appearance closer to their chronological age.

Beighton et al. advocated plastic surgical intervention at an early age in patients with congenital cutis laxa to avoid social anxiety in school-going children and the same can be applied for patients with acquired cutis laxa.

Our patient did not need a rhytidectomy due to the localized affliction of infraorbital regions bilaterally, and excision of the affected portion of skin resulted in an acceptable outcome and was under regular follow-up and was faring well at the end of 1 year following surgery with no signs of recurrence.

In conclusion, plastic surgical procedures, rhytidectomy being the first choice, have a definite role in the management of acquired cutis laxa. Although multiple procedures may be required, the psychosocial benefits need to be taken into consideration, and early surgical intervention is advised.

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 anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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