EPIDERMOLYSIS BULLOSA: CURRENT SURGICAL MANAGEMENT AND REVIEW OF THE LITERATURE

Birol CİVELEK, D. TANYEL, E. BİLGEN, H.İ. İNAL, K. AKSOY, S. ÇELEBİOĞLU

ABSTRACT

Epidermolysis bullosa is a genetically determined chronic disabling disease. The disease is classified in non-scarring or simple and scarring or dystrophic types. Various medical and surgical approaches are reported with no sustainable results.In this report, nine patients treated with split thickness skin grafting and flaps are reported. Due to recurrence of the contractures, a long term benefit is almost always impossible in these patients. However, early surgical interventions and physical therapy may be of help for the progression of the disease.

Keywords: Epidermolysis bullosa, graft, reconstruction

ÖZET

Epidermolizis bülloza, genetik olarak tanımlı, kronik, sakatlık bırakan bir hastalıktır. Hastalık, basit ya da skarsız ve distrofik ya da skarlı olarak sınıflandırılır. Sürdürülebilir sonuçları olmayan medikal ya da cerrahi tedaviler bildirilmiştir. Bu çalışmada, kısmi kalınlıkta deri grefti ya da fleplerle tedavi edilen dokuz vaka bildirilmiştir. Kontraktür rekürrensine bağlı olarak, bu hastalarda uzun süreli yarar sağlamak çoğu zaman olanaksızdır. Bununla birlikte, erken cerrahi müdahale ve fizik tedavi hastalığın seyrinde yararlı olabilir.

Anahtar Kelimeler:Epidermolizis bülloza, greft, rekonstrüksiyon

INTRODUCTION

Epidermolysis bullosa is a genetically determined chronic noninflammatory disease typically characterized by skin blisters following minor trauma. About 20 different types of epidermolysis bullosa have been described.[1-4] Generally, the disease is classified in two categories: nonscarring or simple and scarring or dystrophic types. In the simple type, the blisters form above the basement membrane whereas it is in the upper dermis in the scarring type. It has been shown that dystrophic type is associated with intradermal collagen degeneration and a variable loss of anchoring fibrils in the papillary portion of the dermis.[5-7] The distinction between these two entities is extremely important in the management.

MATERIAL AND METHODS

Nine patients were treated with epidermolysis bullosa related problems. These patients were admitted due to wound problems such as epithelization failure or contractures over the flexion creases. Their age varied from 10 to 16. Five were female and four of the patients were male. They underwent surgery for mitten hands and pseudosyndactyly and contractures over the flexion creases in the hands and the feet and epithelization problems. In the early postoperative period they were put on physical therapy program. They were followed up for a period of three years. In long term, four of these patients had recurrence of syndactyly in their feet and hands. Those recurrent cases underwent revision surgery. Nevertheless, the results were not very satisfactory and they refused further surgical interventions.

Case I

This 7 year old girl presented to the clinics with the complaints of hand deformity and recurring lesions on the legs. She was referred from a rural region where she had been treated with topical wound care and oral antibiotics on multiple occasions. On physical examination, she was found to have mitten hands and non-healing ulcers on the legs. She underwent contracture release and split thickness skin grafting under general anesthesia (Figure 1). Postoperatively, her hands were put on splint and physical therapy. However, she presented one year later with contracture in the flexion creases of the fingers.

Case II

This 14 year old boy presented with contractures in his hands. He underwent release of contractures and reconstruction with local flaps. Two years into his surgery, he presented with recurring contractures. He had a second surgery for release. He was put on pysical therapy and splinting. He is currently complaining of contractures but refusing to undergo further surgery (Figure 2).

Case III

This 17 year old girl presented with severe contractures of his fingers (mitten hand) and toes with nonhealing wounds on arms and legs. She had release of contractures to some extent. However, it was not possible to obtain full extension due to contractures of the joints. She refused further treatment and lost to follow up (Figure 3).

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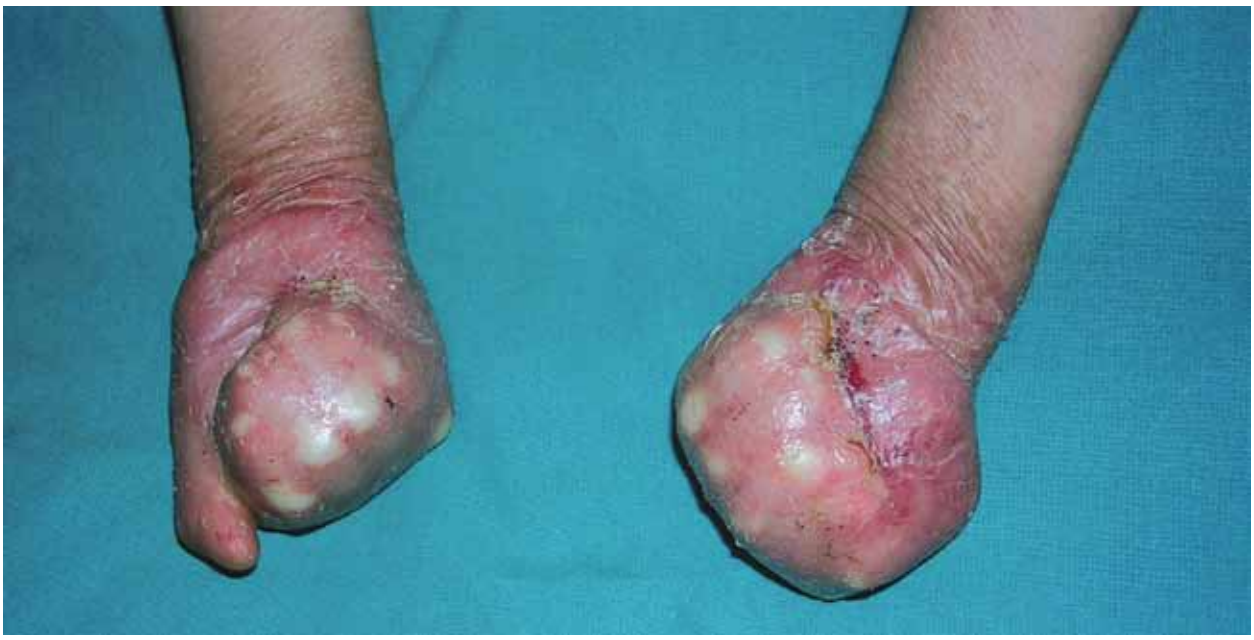


Figure 1a: Mitten hands appearance in this 7 year old girl.

DISCUSSION

Scarring in dystrophic epidermolysis bullosa may result in disabling deformities particularly in the hands. Due to fragile skin and mucosa, these patients pose potential problems even during anesthesia. Treatment modalities of hand and foot deformities in epidermolysis bullosa are reported in various publications.[8-10] Patients with epidermolysis bullosa particularly those with functional disabilities present with difficulties in skin care, surgical management and during anasthesia and postoperatively. It is reported that correction of advanced deformities by simply removing the epidermal cocoon is not sufficient for correction of deformities.[1] The surgical treatment should be aimed for release of contractures over the web spaces and flexion creases with the use of split thickness skin grafts or skin flaps.

The relevant literature lacks a wide coverage of surgical treatment modalities in epidermolysis bullosa. It is noted that surgical interventions do not alter the course of disease. However, the corrections are not sustainable and the recurrences are inevitable. In our series, acute cases of epidermolysis bullosa have been treated as acute burn care. Particularly those with acute inflammation and epidermis loss, they have been managed with split thickness skin grafts or temporary skin dressings have been applied until epithelization occurred.

It seems apparent that acute cases of epidermolysis bullosa need to be approached like a burn wound to prevent bacterial infection and early occurrence of contractures. Those cases with contractures already present, conventional methods of reconstruction such as flaps and skin grafts need to be used. Postoperative splinting of the hands is also of utmost importance to prevent early recurrence of contractures. However,

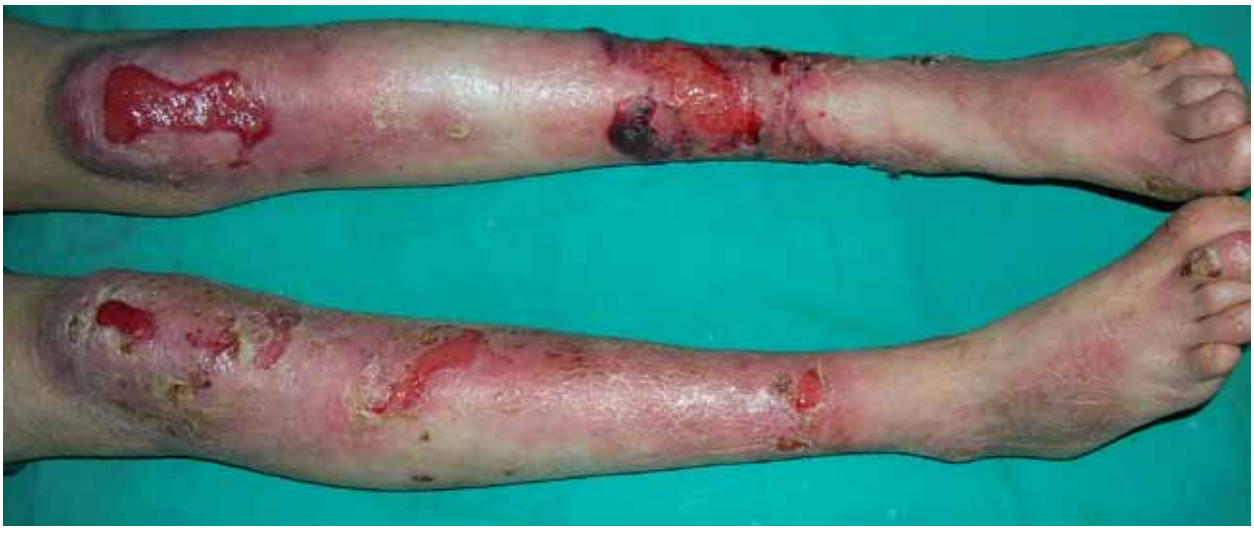
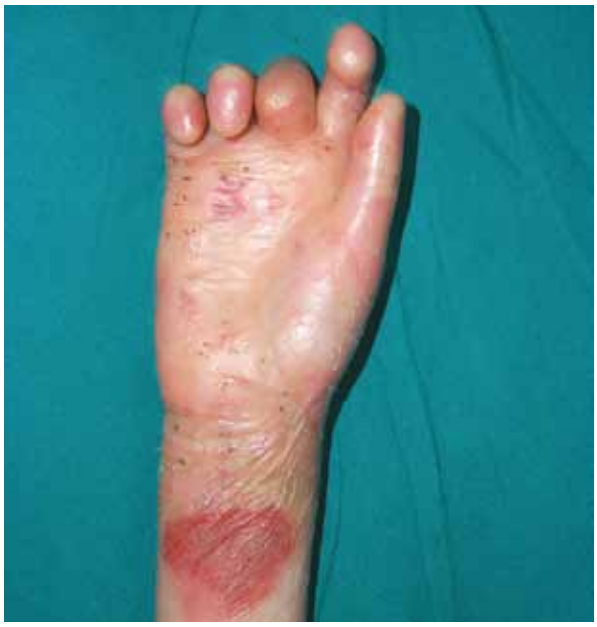


Figure 1b: Nonhealing wounds on the lower extremities in the same patient.



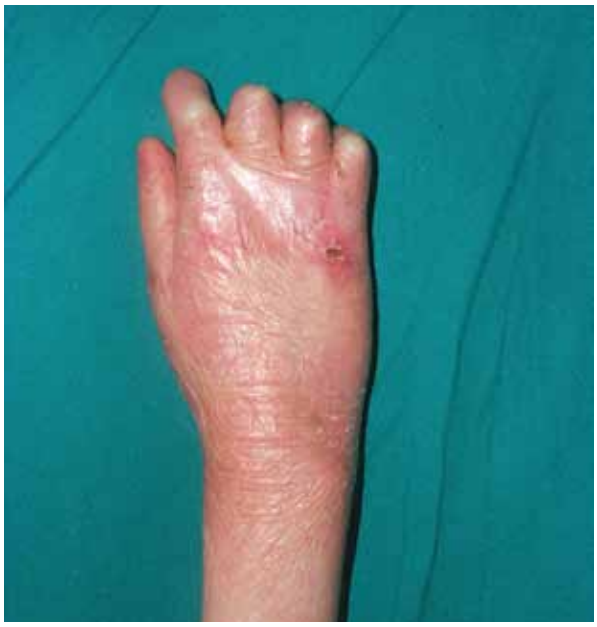


Figure 2a, 2b: Severe contracture of the hand in 17 year old boy.

long term outcomes are not always satisfactory. These patients should be aware of dismal outcomes of minute traumas. Therefore, early surgical treatment in childhood is mandatory to prevent advanced contractures of joints since there is no definitive curative surgical or medical treatment. These patients require close follow up and training for future problems. The role of reconstructive surgery in the management of epidermolysis bullosa should not be dismissed. Plastic Surgery must get involved in the early period of the disease progression.

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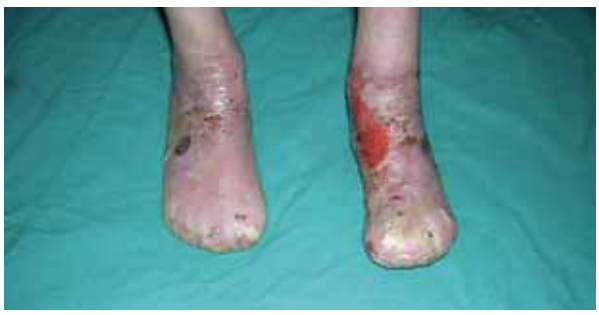

Figure 3: Severe contractures of the web spaces in the feet of a 17 year old girl.

KAYNAKLAR

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