Treatment of Severe Hand Deformities Caused by Epidermolysis Bullosa

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Little research has been done regarding the treatment of severe hand deformities caused by epidermolysis bullosa. A 14-year-old boy was diagnosed with congenital epidermolysis bullosa. He was treated in our hospital several times, but the pathogenetic condition worsened. On examination, both hands were clenched fists and had scar formation. Skin fusion was observed between the 5 fingers. Nails were absent and the thumb was in the fist. His fingers were short, and active and passive flexion and extension could not be performed. The right hand was treated first. After the adhesions were separated, we found that the 5 fingers were connected by dermis. After the dermis was separated and the hand was fixed in the extension position, there were small cutaneous deficiencies. The fingers were fixed in the functional position with Kirschner wires. The wound surface was covered with self-made aureomycin ointment gauze. After regular dressing changes for 6 weeks, the wound surface was completely healed. After 3 months of rehabilitation training, most hand function was recovered.

Based on our findings in this case, when treating patients with epidermolysis bullosa, physicians must carefully observe whether enough dermis exists to avoid an unnecessary skin graft. We also found that the quality of skin used in skin grafting is questionable due to pathological changes in the skin. If there is enough dermis and the cutaneous deficiency is smaller after contracture release, the covering of drug dressings on the cutaneous deficiency is more conducive to the recovery of limb function and the reduction of damage to the donor sites.

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Epidermolysis bullosa is a genetic disease characterized by increased skin fragility. Minor trauma to the skin can lead to blisters. The incidence of autosomal dominant epidermolysis bullosa is 1:50,000, and the incidence of autosomal recessive epidermolysis bullosa is 1:200,000 to 1:500,000 in infants. Currently, no effective therapeutic methods for epidermolysis bullosa exist. Oral glucocorticoids, vitamin E, or phenytoin sodium can relieve pathogenetic conditions. Little research has been done regarding the treatment of severe local lesions caused by epidermolysis bullosa. This article describes a boy with epidermolysis bullosa who had severe hand flexion deformities and obtained improvement in hand function after surgical treatment.

CASE REPORT

A 14-year-old boy had a vesicle on his right foot when he was born and was diagnosed with congenital epidermolysis bullosa in our hospital. He was prescribed glucocorticoids, vitamin E, and phenytoin sodium, but his pathogenetic condition continued to worsen. He was referred to our hospital due to functional loss of both hands in March 2010.

On examination, both hands were clenched fists and had scar formation. There was skin fusion of the 5 fingers. Nails were absent and the thumb was in the fist. The fingers were short, and active and passive flexion and extension could not be performed (Figure 1). We treated the right hand first due to a lack of experience with the treatment of epidermolysis bullosa of the hand.

Anesthesia was performed by brachial plexus block. The 10-layer cotton paper served as a liner, and the tourniquets were placed on the paper around the right upper arm to stop bleeding. After the adhesions were separated, we found that his 5 fingers were connected by dermis. When the dermis was separated and the hand was in placed in the extended position, small cutaneous deficiencies were observed. The thumb was fixed in an opposition position using 3 Kirschner wires (Weigao Company, Shenyang, China) and the other fingers were fixed in a functional position using a Kirschner wire. The patient was prescribed intravenous antibiotics to prevent infections for 1 week postoperatively.

Aureomycin, a highly effective and broad-spectrum antibiotic, has had therapeutic effects on small-area bacterial infection and rhagades of the hands and feet in the winter. Therefore, in this study the small wound surface was covered with self-made aureomycin ointment gauze (Northeast General Pharmaceutical Factory, Shenyang, China) (Figure 2). Regular dressing changes were performed for pin-site infections (Figure 3). After regular dressing changes for 6 weeks, the wound surface had healed completely (Figure 4).

Six weeks postoperatively, the Kirschner wires were removed, and the extension losses of the thumb and fingers were approximately 0.3 and 0.4 cm, respectively (Figure 5A). After wire removal, rehabilitation began. During the 3-month rehabilitation period, splints were used to maintain the finger and thumb positions; dressing changes were performed if infection occurred.

After 3 months of rehabilitation training, range of motion (ROM) of the thumb reached 30° abduction. Palmar opposition and thumb pinch strength were almost normal. Flexion range of the interphalangeal joint of the thumb reached 25°.

Active and passive ROM of the metacarpophalangeal joint of the second to fifth fingers were 45° and 50°, respectively. Active and passive ROM of the proximal interphalangeal joints of the second to fifth fingers were 50° and 60°, respectively (Figure 5B). Active and passive ROM of the distal interphalangeal joints of the second to fifth fingers were 30° and 40°, respectively. The pain and temperature sensations failed to restore.

DISCUSSION

Epidermolysis bullosa, a type of genodermatosis, is characterized by blisters and bulla of the skin due to congenital defects of skin structural protein. According to the location of the dermatolysis as viewed under a transmission electron microscope, epidermolysis bullosa is divided into 3 types: epidermolysis bullosa simplex, junctional epidermolysis bullosa, and dystrophic epidermolysis bullosa. Each type has a different clinical manifestation.
In epidermolysis bullosa simplex, blisters are located in only the basal cell layer of epidermis, skin lesions are mild, and no scars form after healing. Skin lesions only occur at the limb ends and on the extensor side of limb joints and do not invade mucous membrane. When children crawl several months after birth, blisters readily occur in the friction sites.

Junctional epidermolysis bullosa is rarely seen. Children with junctional epidermolysis bullosa have large-area blisters, bulla, and erosion in the skin after birth, and generally die within 2 years of birth.

In dystrophic epidermolysis bullosa, blisters are under the lamina densa of the basilar membrane, and skin lesions are severe, with scar formation after healing. Skin lesions can occur on any part of the body surface, including mucous membranes, but are especially prone to occur at limb ends. Repeated blisters and scarring of the limb ends lead to skin adhesions between the fingers (or toes) and digital bone atrophy, which manifests clawed or clenched fists. Repeated oropharyngeal mucosal diabrosis and scarring lead to difficulty opening the mouth and dysphagia. Children with dystrophic epidermolysis bullosa may have skin lesions after birth, and the prognosis is poor.

In the current patient, both hands were clenched fists and syndactyly with complete loss of activity was observed. He was diagnosed with dystrophic epidermolysis bullosa. No effective and radical method exists for the treatment of dystrophic epidermolysis bullosa. Currently, the treatments for dystrophic epidermolysis bullosa are mainly preventive and supportive, including nutrition support, anemia correction, avoidance of trauma, infection prevention, and oral high-dose vitamin E. In patients with severe skin and mucous membrane injury, glucocorticoids and phenytoin sodium may be given to reduce scar formation.3-5

For the treatment of severe hand deformities caused by epidermolysis bullosa, Ladd et al6 used a free skin graft to cover the cutaneous deficiency after contracture release. However, 2 problems occur when a free skin graft is used for patients with epidermolysis bullosa. First, if the patient’s skin is used, its quality is questionable because there are pathological changes in epidermolysis bullosa skin. This skin may have adverse effects on skin graft survival and healing. Second, the skin quality of patients with epidermolysis bullosa is poor. Minor trauma can lead to blisters, and then scars occur after blister healing. Although the skin grafts survive, a large scar area is unavoidable, and the scar at donor sites is also severe. Therefore, we should be cautious about free skin graft in patients with epidermolysis bullosa.

We decided to treat the right hand first. After accumulating some experience, we then treated the left hand. Fortunately, although there was epidermis deficiency, dermis was still present. All adhesions belonged to dermis pseudo-healing status. After the skin and subcutaneous tissue were separated and the hand was placed in the extension position, there was only small cutaneous deficiency that did not require skin grafting.

Dressing changes were necessary because there was a large portion of exposed dermis and small cutaneous deficiencies in fingers and palmar side. To avoid the adhesions between the dressings and the wound surface and to prevent infection, we used self-made aureomycin ointment gauze to cover the wound surface. The dressing change was performed by the surgical staff once daily because changing dressings too frequent affects skin healing. Because the dressings have an anti-adhesion effect, the dressing change is relatively tolerable to the patient.

Aureomycin is used to fight an infection of gram-positive cocci (especially Staphylococcus and Pneumococcus), rickettsia, mycoplasma, chlamydia, and atypical mycobacteria. Aureomycin can inhibit bacterial protein synthesis by linking to the bacterial ribosomal 30S subunit. Au-
reomycin is mostly for local application, with little absorption and rare adverse reactions. In clinical practice, we have found that aureomycin has better therapeutic effects on rashes on the hands and feet, which is associated with its anti-infection and anti-adhesion. Whether aureomycin has the promotive effect on wound healing remains to be further explored.

The wound surface completely healed after the 6-week dressing change. It is worth noting that to avoid skin damage caused by the tourniquets, they were placed on the 10-layer cotton paper around the right upper arm to stop bleeding. Postoperative results have proven to be effective.

Based on our findings from this case, patients with epidermolysis bullosa must be carefully observed to determine whether enough dermis is available to avoid an unnecessary skin graft and whether the quality of epidermolysis bullosa skin used in skin grafting is acceptable because there are pathological changes in epidermolysis bullosa skin. If there is enough dermis and the cutaneous deficiency is smaller after contracture release, using dressings with an antibiotic on the cutaneous deficiency is more conducive to the recovery of limb function and the reduction of damage to the donor sites.

REFERENCES