Hailey-Hailey Disease Successfully Treated with Carbon Dioxide Laser
-A Case Report

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A 45-year-old man presented with erythematous vesicles and erosions on erythematous backgrounds on his axillae, groin, and popliteal fossae. He had been treated with corticosteroid ointments for several years, but the clinical course waxed and waned, and the lesions never completely disappeared. A skin biopsy specimen from his left axilla revealed suprabasal acantholysis (a "dilapidated brick wall"). Diagnosis of recurrent Hailey-Hailey disease was made after careful analysis of the clinical and histopathological features. We administered a CO2 laser to eradicate the unresponsive remnants of the lesion. The operative site re-epithelialized within 3 weeks. One year after the treatment, there was no evidence of recurrence. For recalcitrant cases of Hailey-Hailey disease, as in this case, the use of a CO2 laser may be an effective therapeutic modality. (Dermatol Sinica 25: 132-135, 2007)

Key words: Hailey-Hailey disease, Familial benign pemphigus, CO2 laser

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Accepted for publication: December 22, 2006
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INTRODUCTION

Hailey-Hailey disease, or familial benign pemphigus (FBP), is a rare blistering disorder first described by the Hailey brothers in 1939.1 It is characterized by recurrent vesicles and erosions, particularly involving flexural areas. The major underlying pathologic process in Hailey-Hailey disease (HHD) is acantholysis, and the changes appear in isolated keratinocyte cultures, indicating a fundamental defect in the keratinocytes themselves.2 HHD is an autosomal dominant disease. The causative genetic defect was identified as a mutation in ATP2C1, the gene on chromosome 3q21-24 coding an ATP-driven transmembrane calcium pump.3-6 A variety of medical and surgical therapies exist for HHD. We report a recalcitrant case with typical HHD erythematous vesicles and erosions on an erythematous background on the axillae, groin, and popliteal fossae which was effectively treated with CO2 laser ablation.

CASE REPORT

A 45-year-old man had erythematous vesicles and erosions on an erythematous background on his axillae, groin, and popliteal fossae for about 10 years (Fig. 1a). His brother also had similar skin lesions. The lesions were precipitated by friction and excessive sweating, and an unpleasant odor arose from the diseased areas. He had been treated with corticosteroid ointments for several years, but the clinical course waxed and waned, and the lesions never completely disappeared. Skin biopsy specimens from the erythema on his left axilla and groin revealed parakeratosis, hyperkeratosis, and suprabasal acantholysis (Fig. 2). From these clinical and histopathological findings, he was diagnosed as having HHD, or FBP. Laboratory findings including hematological parameters, renal and liver functions were all within normal limits. Culture of the exudate from a skin lesion in his groin revealed the growth of Candida albicans, Proteus mirabilis, Viridans streptococci, and β-streptococci group B species. The patient was treated with oral antibiotics (minocycline HCl at 100 mg twice a day), a topical antifungal agent: exelderm (Sulconazole) cream, and topical antibiotics: fusidate cream.

Because of his refractory disease, he decided to pursue CO2 laser resurfacing of his
axillae. After disinfection of the lesional sites and infiltration anesthesia with 1% Xylocaine and 1:100,000 epinephrine, continuous CO₂ laser vaporization was performed with three passes over the affected axillary tissue using a Truespot 3-mm collimated handpiece (Lumenis Compact 30C CO₂ Laser). The power setting was 3.0 W to ablate all of the epidermis. The surfaces were wiped between the passes. Postoperatively, the patient was instructed to apply topical antibiotics with fusidate cream to the wounds until they healed. The wound healing from the CO₂ laser treatment in our patient was complete in 3 weeks. Besides, the patient applied topical antifungal to his skin lesions. About 1 year later, the patient had a flare-up of disease involving his groin, buttocks, and left popliteal fossa (Fig. 3a). However, the treated areas were strikingly spared (Fig. 1b).

Encouraged by these results, we used the same procedures to treat his left popliteal fossa. Recurrence of the disease was noted about 18 months later (Fig. 3b), however, all of the treated areas have shown resistance to repeated involvement with HHD.

DISCUSSION

HHD is a rare autosomal dominant genodermatosis. It presents with recurrent vesicles and erythematous crusting erosions in areas subjected to friction, including the neck and intertriginous areas. Lesions are frequently precipitated by friction and infection with various bacteria.

Various therapeutic modalities have been used in HHD. The disease often responds to topical steroids with antibacterial agents, either local or systemic. Other useful therapies include dapsone, methotrexate, thalidomide, ciclosporin, and psoralen plus ultraviolet A. While medical therapies may temporarily suppress the disease, recurrences are common, and patients often seek a more definitive treatment. Surgical intervention may be required to achieve prolonged remission or a complete cure. Full-thickness excision with or without split-thickness grafting, dermabrasion, electrodessication, cryosurgery, and reduction mammoplasty have all been advocated for recalcitrant disease.

An alternative to surgical intervention is ablation with a CO₂ laser. The CO₂ laser is a far-infrared laser with a wavelength of 10,600 nm, emitted when carbon dioxide molecules are excited using DC currents. CO₂ laser ablation is a safer, low-bleeding method with less postoperative pain than dermabrasion, which is followed by rapid healing of the erosions. The CO₂ laser operates by rapidly heating the cells and vaporizing the intracellular water, causing cell destruction and degeneration of the tissue. The use of a CO₂ laser for dermatological procedures offers a number of advantages: first, curettage can be performed within the designated range and depth; second, the target tissue can be vaporized while leaving the unaffected areas intact by surrounding the lesion with wet gauze which cools the temperature of the perilesional tissue; and lastly, the procedure is rapid and blood loss is minimal. Kartamaa and Reitamo described the use of a continuous CO₂ laser in six patients with symmetrical lesions of HHD, using one side as an untreated control. Laser vaporization was performed on the treated side until healthy dermis was seen; of the six patients, five noted significant improvement in the laser-
treated lesions compared to the untreated control sites at follow-up (which ranged 4~27 months). Hypertrophic scarring occurred in the axilla of one patient.12

The mechanism of durable healing of HHD lesions after vaporization of the affected areas with the CO₂ laser resurfacing or dermabrasion is unclear. Theories maintain that the superficial erosions are re-epithelialized by rapidly proliferating adnexal keratinocytes that do not express the molecular defect of HHD.8, 13-15

In summary, we elected to perform CO₂ laser therapy on the grounds that it was the least-invasive method of surgical intervention with minimal scarring. Our patient has clearly benefited from laser resurfacing of an area prone to involvement with HHD. This report supports previous studies suggesting that for severe cases of Hailey-Hailey disease, ablation of affected areas with modern CO₂ laser systems may be the surgical treatment modality of choice. However, further studies are needed to confirm these results and to determine the long-term effects of this treatment and the duration of remission.

REFERENCES