Efficacy of Erbium:YAG Laser Ablation in Darier Disease and Hailey-Hailey Disease

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Background: Among different surgical approaches, dermabrasion and carbon dioxide laser vaporization have been used to treat Hailey-Hailey disease (HHD) (familial benign chronic pemphigus) and Darier disease (DD) (keratosis follicularis), with various results. Because of the erbium:YAG laser’s unique absorption characteristics in tissue water, erbium:YAG laser ablation combines the advantages of both techniques, avoiding thermal injury of vaporization and also allowing selectively deeper tissue removal in the follicular lesions of DD. Therefore, good results should be expected in both types of acantholytic disorders.

Observations: Four patients (2 with HHD and 2 with DD) with different affected areas were treated with laser ablation. During a follow-up period ranging from 8 to 20 months, complete remission was achieved in 3 patients—2 with DD and 1 with HHD—and significant improvement was achieved in 1 patient with HHD. Histological examination of control biopsy specimens after ablation in 1 patient with DD revealed no signs of the disease and only a slight fibrosis in the papillary dermis.

Conclusions: Erbium:YAG laser ablation effectively removes lesions of both HHD and DD and can also yield excellent long-term results in chronic, recalcitrant cases.

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DARIER DISEASE (DD) (keratosis follicularis) and Hailey-Hailey disease (HHD) (familial benign chronic pemphigus) are autosomal dominant disorders characterized by specific alterations corresponding to seborrheic areas of the body and intertriginous areas.1-3 The histological features consist of dyskeratosis, suprabasal acantholysis, papillomatosis, and suprabasal separation with extensive loss of intercellular bridges.4 Despite the clinical and histological overlap of the 2 diseases, the underlying mutations of both affect different genes.5

Conventional therapeutic approaches consist of topical and systemic steroids, antibiotic agents, vitamin A acid cream, and oral retinoids. Nevertheless, prolonged remission is difficult to achieve.1-3

Good results for both diseases were obtained from such surgical interventions as excision6-7 or excision with subsequent split-thickness skin grafting,8 electrodesiccation,9 and dermabrasion.10-14 According to the observations of other authors, our own experiences have confirmed the value of dermabrasion, particularly in the treatment of HHD. However, DD frequently requires selective removal of keratotic papules and circumscribed deeper ablation in follicular lesions, which is not feasible with dermabrasion. Moreover, the treatment of certain areas, such as the upper trunk area or the neck, is limited because of the risk of scar formation.

Recent case reports have advocated the use of carbon dioxide laser vaporization in the treatment of HHD and DD,15-17 a procedure that could compensate for some of the disadvantages of dermabrasion. However, carbon dioxide laser vaporization, even in pulsed mode, leaves residual thermal damage, especially with deeper tissue removal, and therefore increases the risk of scar formation and prolonged wound healing. In contrast, erbium:YAG laser ablation, operating at a wavelength of 2.94 µm, combines the most efficacious stepwise etching of the skin surface with the least thermal damage owing to its unique absorption characteristics in tissue water.18,19 This technique should therefore allow the precisely controlled removal of lesions in both HHD and DD and limit the injury in widespread lesions or in those located on areas prone to keloid formation. We decided to treat 4 patients (2 with HHD and 2 with DD) with pulsed 2.94-µm erbium:YAG laser ablation and...
TECHNIQUE

After written consent was obtained, erbium:YAG laser ablation was performed with the patients under local anesthesia. To ensure that the expected improvement was the result of the surgical intervention and not attributable to the spontaneous course of the disease, untreated control lesions were left in all 4 patients before complete ablation was performed. After a minimum follow-up of 8 weeks and a successful treatment outcome, the residual areas were treated in further sessions.

Surgery was performed with an erbium:YAG laser (MCL 29 Dermablate; Aesculap Meditec GmbH, Jena, Germany). Depending on the lesions, we used spot sizes varying from 1.6 mm for small papules of DD to 5.0 mm for confluent plaques of HHD, with repetition rates of 5 to 10 Hz and energies between 300 and 1000 mJ, resulting in fluences of 5 to 8.5 J/cm2 per pulse (Table). The pulse width used in all treatment sessions was 350 microseconds.

Large areas were treated with the painting technique, which implies an overlap of approximately 30%. Patients 1 and 2 exhibited numerous punctate lesions, which were treated with a point-by-point technique. According to our previous experiences, and in agreement with the observations of others, we also treated a margin of adjacent uninvolved skin. Up to 7 stacked passes (pulse series) per lesion were performed, and the resulting debris was gently removed with saline-soaked gauze.

The clinical end point of the ablation procedure was determined by the disappearance of the brownish papules of DD and the grayish red lesions of HHD and by the appearance of a white tissue layer exhibiting papillary vessels and pinpoint bleeding after several ablation passes. Ablation of all lesions was performed until the papillary dermis was exposed and a slight punctate bleeding was induced.

Postoperatively, the wounds were covered with a nonadhesive dressing (Cuticerin; Beiersdorf AG, Hamburg, Germany) and sulfadiazine-silver cream. The patients did not receive systemic or topical antibiotics. The dressings were used for approximately 10 to 12 days, depending on the degree of reepithelialization. Dressings were changed every day and cleansed with saline-soaked gauze. In contrast to resurfacing procedures for other indications, corticosteroid formulations were not applied to exclude the possibility that any improvement was due to their anti-inflammatory effects.

Four patients (2 men and 2 women) with characteristic clinical and histological lesions of chronic recalcitrant DD and HHD were included in our study. The medical history of all 4 patients revealed that they had received topical treatment with corticosteroids and/or antimicrobial agents but that complete remission had not been achieved in any of them.

CASE 1

A 57-year-old woman presented with a 12-year history of recurrent, pruritic, unilateral, zosteriform, erythematous papules in the right axilla, scalpular region, and upper arm area. The histological findings in multiple biopsy specimens were consistent with the clinical diagnosis of DD.

CASE 2

A 56-year-old woman presented with multiple hyperkeratotic, brownish red papules that involved the trunk and extremities and coalesced to wartlike plaques on the flexural sites and the neck (Figure 1). She complained of severe pruritus, which had been resistant to topical treatment that was initiated 2 years ago, when the disease had worsened. Histological examination of the lesions confirmed the diagnosis of DD.

CASE 3

A 47-year-old man presented with a 3-year history of HHD involving the groins, the lateral aspect of the scrotum, and both axillae. He was severely distressed by the malodor of the lesions. Skin biopsy specimens showed the characteristic histological features of familial benign chronic pemphigus.

CASE 4

A 42-year-old man presented with a 3-year history of HHD that affected the groins and both axillae. His family had a history of the disease, and the histological findings were consistent with the diagnosis. Pain, pruritus, and excoriation of the lesions, as well as their chronicity, led to unemployment and social retirement.

RESULTS

Laser test ablations were successful in all patients and showed complete regression within the ablated areas, whereas the untreated sites were still affected. In patients 1, 2, and 3, complementary laser removal of control lesions led to excellent results (Figure 2), and the treated areas showed no recurrence during follow-up (Table). No adverse effects (eg, wound infection, hyperpigmentation, or scarring), other than slight hypopigmentation of the cubital fossa and the popliteal cavity in patient 2, and a few atrophic hypopigmented spots after deeper ablation in patient 1, occurred during the period of observation. It is noteworthy that even the plaquelike areas on the ventral aspect of the neck of patient 2 could be cleared completely without any adverse effects even though this region is at high risk for scar formation (Figure 2, right). Both patients with DD also experienced complete relief from itching. It was interesting to observe that ablation of solitary follicular hyperkeratotic papules in the patients with DD resulted in a “wicklike” situs rep-
resented by a small prominent keratotic core in the center of the lesion and needed additional focused single pulses for removal. This “wick phenomenon” is likely to be caused by reduced water content (absorbing target) and consequently by less effective ablation within the central hyperkeratotic follicular area.

In patient 3, who had HHD, a complete remission of the affected sites was achieved after ablation and no disease activity was detected during more than 1 year of follow-up, whereas in patient 4, some areas recurred at the edges and adjacent to the treated sites, requiring an additional session.

In all 4 patients, the clinical diagnosis was confirmed by histological examination, which revealed the characteristic findings of DD and HHD (Figure 3). Biopsy specimens were obtained from patient 2 immediately after laser ablation and showed the removal of the entire epidermis and stratum papillare, preserving adnexal structures (Figure 4). Only minimal signs of thermal damage were present in the underlying dermis. Eighteen months after the laser procedure, control biopsy specimens were obtained from the treatment site of patient 2. No signs of DD could be detected by histological examination, and the papillary dermis exhibited only a slight increase of fibrotic tissue (Figure 5).

Eight months after laser ablation, the control biopsy specimens from patient 1 revealed the same results as those from patient 2. Unfortunately, both patients with HHD refused to undergo additional biopsies.

COMMENT

It has been shown by several authors that surgical removal of lesions can achieve successful treatment results in both HHD and DD. Since a complete excision of the involved skin area is not possible in the majority of cases, a variety of shaving and destructive techniques has been introduced, with varying outcome, since each technique has its specific limits, depending on the individual prerequisites of a given lesion. Recent progress in pulsed laser resurfacing procedures has led to the development of more refined techniques that allow highly controlled, selective ablation of the skin, even on critical sites of the body surface where dermabrasion might be impossible or where thermal vaporization could induce

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Treatment Sites</th>
<th>Fluence, J/cm²</th>
<th>Maximum No. of Pulse Series</th>
<th>Results</th>
<th>Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Axilla, scapular, and upper arm region</td>
<td>7.1</td>
<td>6</td>
<td>Complete remission</td>
<td>11</td>
</tr>
<tr>
<td>2</td>
<td>Trunk, extremities, ventral aspect of neck</td>
<td>5.0</td>
<td>7</td>
<td>Complete remission</td>
<td>20</td>
</tr>
<tr>
<td>3</td>
<td>Axillae, groins, scrotum</td>
<td>8.5</td>
<td>5</td>
<td>Complete remission</td>
<td>15</td>
</tr>
<tr>
<td>4</td>
<td>Axillae, groins</td>
<td>7.1</td>
<td>5</td>
<td>Partial remission</td>
<td>8</td>
</tr>
</tbody>
</table>
unwarranted tissue trauma and interfere with wound healing. In experimental and clinical work, pulsed skin ablation at a laser wavelength of 2.94 µm (absorption peak of tissue water) has been shown to combine a stepwise char-free ablation with the possibility of producing circumscribed deeper crater lesions without cumulative coagulation, even after serial pulses.

Our experience in patients with HHD and DD has confirmed that these advantages are of special value in treating thicker recalcitrant lesions, for which deeper ablation is required, as well as in treating papular lesions located on critical sites. Carbon dioxide laser removal, in contrast, even in pulsed modes, can produce cumulative necrosis after serial pulsing (usually more than 3 passes) owing to tissue desiccation with loss of absorbing water on the irradiated surface.18,19

Therefore, thermal laser vaporization and electrodesiccation are limited in deeper lesions, where a risk of hypertrophic scar or keloid formation has to be considered. We did not observe the development of hypertrophic scars in our patients, which was reported to occur after carbon dioxide laser vaporization in a patient with familial benign chronic pemphigus.17

On the other hand, dermabrasion is unsatisfactory in treating papular lesions and in critical sites, and therefore remains of restricted value in most cases that present with typical distribution patterns of DD. To our knowledge, there have been only 2 reports of patients with
DD who were treated with laser surgery. In contrast to those 2 cases, we did not administer any concomitant therapy that might influence the spontaneous course of the disease.

Long-term follow-up showed excellent results, with complete remission in 3 patients—2 with DD and 1 with HHD—and partial remission in 1 patient with HHD. The exact mechanism of how the removal of lesional skin contributes to the therapeutic success is as yet unknown. We do not agree with the view of other authors that necrosis and the subsequent fibrosis of the papillary dermis represent a prerequisite for improvement.

It is more likely that a critical depth of amount of tissue removal is essential and does not necessarily represent tissue necrosis. Our histological findings showed only minimal thermal damage after ablation and slight fibrosis of the papillary dermis after wound healing in ablation craters of DD. Therefore, we were even encouraged to treat the ventral neck area of 1 patient, and complete remission was achieved without any adverse effects (Figure 2, right). The clinical results, as well as the histological findings, confirm that erbium:YAG laser ablation is highly controllable owing to an extremely low ablation threshold, which enables a procedure to be carried out in “micrometer steps.” Therefore, the ablation depth corresponds well to the overall amount of tissue destruction, since the relative amount of additional underlying damage is negligible. This result is in contrast to the outcome that is seen with superficial carbon dioxide laser vaporization, where cumulative damage due to tissue heating has to be carefully considered, especially with deeper lesions and additional laser pulses.

In our opinion, erbium:YAG laser ablation represents a very effective and safe therapeutic modality for the treatment of HHD and DD. Fast ablation of large areas, as well as point-by-point removal of keratotic papules and localized deeper ablation, if necessary, is another advantage of the erbium:YAG laser that contributes to the superiority of the method in comparison with abrasive methods or carbon dioxide laser vaporization.

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REFERENCES