

Treatment of the Hailey-Hailey disease with fractional CO₂ laser: a three-case series

Tratamento da doença de Hailey-Hailey com laser de CO₂ fracionado: uma série de três casos

DOI: <http://dx.doi.org/10.5935/scd1984-8773.201791895>

RESUMO

A doença de Hailey-Hailey ou pênfigo familiar benigno é condição rara, que se caracteriza por lesões vesiculares e erosões, associadas a dor e queimação, que comprometem a qualidade de vida dos pacientes. Existem vários tratamentos tópicos e sistêmicos que podem promover temporariamente a remissão das lesões, não existindo tratamento curativo. Algumas opções de tratamento com resultados duradouros abrangem a dermoabrasão e a vaporização com laser de Erbium YAG ou CO₂. Relatamos três casos de pacientes com lesões recorrentes e respostas limitadas aos tratamentos clássicos, que apresentaram melhora clínica importante e alívio sintomático após terapia com laser de CO₂ fracionado.

Palavras-chave: pênfigo familiar benigno; terapia a laser; dióxido de carbono

ABSTRACT

The Hailey-Hailey disease or familial benign pemphigus is a rare condition, characterized by vesicular lesions and erosions with a predilection for intertriginous areas associated with pain and burning sensation that affect the quality of life of patients. There are many topical and systemic treatments for the injuries that can temporarily promote partial or complete remission, but there is no curative treatment. Some treatment options with lasting results include dermabrasion and Erbium laser resurfacing (YAG or CO₂). We report three cases of patients with recurrent lesions and limited responses to classical treatments, which showed significant clinical improvement after fractional CO₂ laser therapy.

Keywords: pemphigus, benign familial; laser therapy; carbon dioxide

INTRODUCTION

The Hailey-Hailey disease or familial benign pemphigus is a rare condition that affects the keratinocytes' adhesion. It has a dominant autosomal inheritance and is characterized by vesicular lesions and erosions, with a predilection for intertriginous areas, such as the armpits, groins, and inframammary region.¹ The lesions appear around the second and third decades of life and can manifest up to the fifth decade.^{1,2}

Its has an indeterminate course, usually with periods of outbreaks and remissions. Eruptions can be triggered by factors such as friction, sweating, heat, emotional stress and ultraviolet radiation. The lesions may have spontaneous remission or worsen with painful fissures or bad odor vegetating lesions, compromising the quality of life in a significant manner.^{1,2}

Several therapeutic options have been described, the most common being antibiotic therapy and oral or topical cor-

Case Reports

Authors:

Vanessa da Nobrega Vilela¹
 Catarina Gonçalves da Silva Carvalho¹
 Gustavo de Sá Menezes Carvalho²
 Angela Cristina Rapela Medeiros³
 Valter Kozmhinsky⁴
 Emmanuel Rodrigues França⁵

¹ Collaborating dermatologist physician, Universidade de Pernambuco (UPE) - Recife (PE), Brazil.

² Medicine student, UPE.

³ Associate Professor, UPE.

⁴ Dermatology Professor, University Hospital Oswaldo Cruz, UPE. Head of the Dermatology Service, Instituto de Medicina Integral Prof. Fernando Figueira (Imip) - Caruaru (PE), Brazil.

⁵ Associate Professor and Head of the Dermatology Service, Faculdade de Ciências Médicas de Pernambuco, UPE.

Correspondence:

Vanessa da Nóbrega Vilela
 Rua Arnóbio Marquês 310, Santo Amaro
 Cep 50100-130 - Recife - PE, Brazil
 Email: van_medufrn@yahoo.com.br

Received on: 28/08/2016

Approved on: 28/02/2017

This study was carried out at the University Hospital Oswaldo Cruz, Universidade de Pernambuco (UPE) - Recife (SP), Brazil.

Financial support: none

Conflict of interests: none

ticosteroid therapy, with varying remission and recurrence rates. Retinoids, systemic methotrexate and cyclosporine, topical tacrolimus, botulinum toxin and photodynamic therapy also led to variable rates of therapeutic success.² The long-term treatment options comprise surgical excision associated with cutaneous graft, and dermabrasion. These therapies yield good results, however with significant rates of complications (i.e. infections and retractions) due to their more invasive character. In contrast, laser therapy is less invasive and has been reported as safe and effective, leading to long-lasting results.³

The authors of the present paper describe three cases of patients who had limited response to classic treatments, nevertheless showed significant clinical improvement, symptomatic relief and absence of recurrence after fractional CO₂ laser therapy.

METHODS

The procedures were performed in an ambulatorial setting, under topical anesthesia (7% lidocaine + 7% tetracaine) (Pliaglis® Galderma, São Paulo, Brazil). The laser device used was the DUAL DEEP® (Lutronics - South Korea), set at the static mode, with a 12mm spot and fluence between 80 and 100 Joules/cm². Two passes (exceeding by 1cm the lesion's visibly active borders) were performed in each of the sessions.

In all cases, it was possible to observe hyperpigmentation sequelae resulting from chronic lesions.

Case reports

Case 1

A 58-year-old female patient had a 6-year history of recurrent Hailey-Hailey disease. She had fissured, hyperkeratotic painful plaques on the dorsum and cervical region. The patient had previously undergone conservative treatment with topical corticosteroids and oral antibiotics, without significant improvement. Multiple areas of the dorsum were treated – one per session – with an average of four sessions in each of the affected areas, over a total period of two years (Figure 1). During a four-year clinical follow up period, the patient did not show recurrence of the lesions in the treated areas.

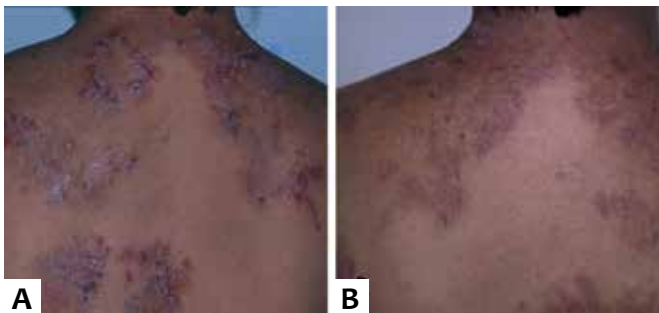


FIGURE 1: A – Pre-treatment appearance; B – Two months after the end of the treatment

Case 2

A 39-year-old female patient had experienced recurrent Hailey-Hailey disease in the axillae for about 10 years. Antimicrobial therapy and oral isotretinoin had been ineffective. Three CO₂ laser sessions were performed in each axilla with monthly intervals (Figure 2). The patient did not present recurrence after 2 years of clinical follow-up.

Case 3

A 46-year-old male patient had been diagnosed with the Hailey-Hailey disease in the adolescence (predominantly in the axillae). There were reports of partial improvement of the lesions with oral antibiotics and topical corticosteroids, however with frequent recurrence. The axillae were then treated monthly with CO₂ laser sessions, in a total of four sessions in each axilla. There was a significant improvement in the lesions, without recurrence after a one year follow-up (Figure 3).

DISCUSSION

Familial benign pemphigus is a disease that courses with recalcitrant erosive plaques and can be debilitating. First-line therapies are generally medicament-based and only promote temporary suppression of the lesions.¹ Dermabrasion and ablative laser, which have been resulting in the long-term remission of the condition, are the most advanced techniques for the treatment of the Hailey-Hailey disease. Dermabrasion is effective, nonetheless limited due to its complications and the impossibility of being performed in certain body sites.²

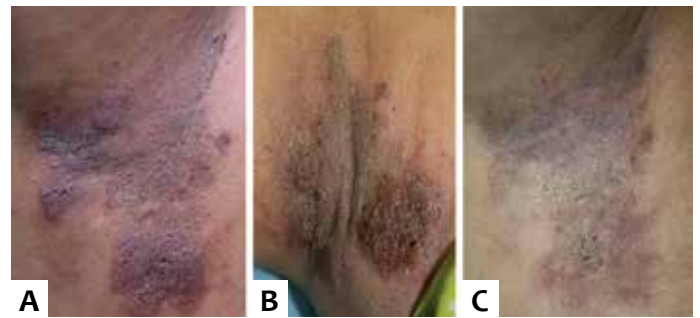


FIGURE 2: A – Before treatment; B – Immediate post-treatment; C – Two months after the treatment

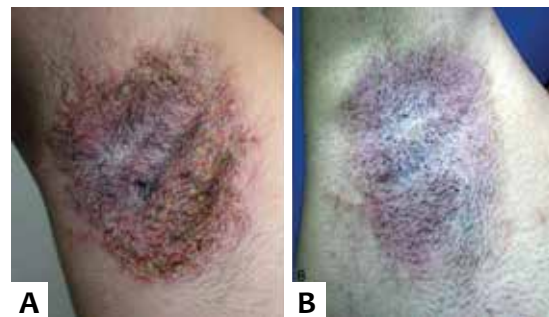


FIGURE 3: A – Pre-treatment appearance; B – Two months after the end of the treatment

On the other hand, laser therapy has several advantages over other therapies: it can be used to treat a large area in a short period of time; it causes less intraoperative pain and less postoperative bleeding; it offers better control of the procedure's depth and decreased probability of scarring as compared to dermabrasion; and it can be performed ambulatorially under local anesthesia.^{2,3} Therefore, it is a promising therapy with the possibility of becoming the method of choice for the treatment of recurrent Hailey-Hailey disease.

The main side effects of CO₂ laser are transient edema and erythema, scarring and depigmentation. A common problem is the recurrence of the disease at the treated lesion's periphery. In order to avoid this situation, Kruppa et al. have suggested that the application of laser should cover an area that exceeds that of the clinically visible disease.^{4,5}

The exact mechanism of action of the ablative laser in this pathology remains uncertain. One theory is that the epidermis and keratinocytes expressing the molecular defect are destroyed, leaving the adnexa intact, allowing the regeneration of the normal epidermis without the adhesion defect. The adnexa have the mutation however they do not express them, being therefore unaffected by the acantholytic process.⁵ Another theory is that dermal fibrosis leads to better support of the unhealthy epidermis and decreases the risk of ulceration and fissure formation.⁶

The three cases reported in the present paper had an excellent response to fractional CO₂ laser, with minimal side effects (erythema, edema and transient pain). They were followed for up to four years with absence of recurrence of the lesions in the treated sites.

Evidence describing the treatment of familial benign pemphigus with CO₂ laser is encouraging in cases such as those described here. Nevertheless, more encompassing studies are necessary to consolidate the status of this method in the therapeutic armamentarium available to treat the Hailey-Hailey disease. ●

REFERENCES

1. Chiaravalloti A, Payette M. Hailey-Hailey disease and review of management. *J Drugs Dermatol*. 2014;13(10):1254-1257.
2. Ortiz AE, Zachary CB. Laser therapy for Hailey-Hailey disease: review of the literature and a case report. *Dermatol Reports*. 2011;3(2):e28.
3. Pretel-Irazabal M, Lera-Imbuluzqueta JM, España-Alonso A. Carbon dioxide laser treatment in Hailey-Hailey disease: a series of 8 patients. *Actas Dermosifiliogr*. 2013; 104(4):325-333.
4. Collet Villette AM, Richard MA, Fourquet F, Monestier S, Gaudy C, Bonerandi JJ, Grob JJ. Treatment of Hailey-Hailey Disease With Carbon Dioxide Laser Vaporization. *Ann Dermatol Venereol*. 2005;132(8-9 Pt 1):637-640.
5. Falto-Aizpurua LA, Griffith RD, Yazdani Abyaneh MA, Nouri K. Laser therapy for the treatment of Hailey-Hailey disease: a systematic review with focus on carbon dioxide laser resurfacing. *J Eur Acad Dermatol Venereol*. 2015; 29(6):1045-1052.
6. Grönemeyer LL, Thoms KM, Bertsch HP, Hofmann L, Schön MP, Haenssle HA. Reflectance confocal microscopy and Hailey-Hailey disease: assessment of response to treatment after CO₂ laser ablation. *J Dtsch Dermatol Ges*. 2014;12(12): 1135-1137.