

Management of Hailey-Hailey disease with Castellani paint



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INTRODUCTION

Hailey-Hailey disease (HHD), also called *familial benign chronic pemphigus*, is a rare genodermatosis first described by the Hailey brothers in 1939.¹ HHD is an autosomal-dominant keratinocyte adhesion disorder secondary to loss of function mutations of the *ATP2C1* gene, an ATPase responsible for calcium homeostasis.² HHD manifests as chronic and recurrent red plaques involving intertriginous sites. Affected areas develop painful vesicles, erosions, fissures, and maceration and often have bacterial, fungal, or viral superinfection. Few treatments are effective for HHD. We report successful treatment of this challenging disease with Castellani paint therapy.

CASE REPORT

A 47-year-old woman presented with a long-standing history of HHD. She was unable to perform many activities of daily living because of substantial pain and pruritus. Prior failed treatments included topical and oral corticosteroids, topical and oral antibiotics, acitretin, azathioprine, etanercept, cyclosporine, radiation, and fractional photothermolysis.

On examination, the woman had pink, macerated, and eroded plaques involving the axilla, inframammary region, and perineum (Fig 1). Histopathologic evaluation found hyperkeratosis, acanthosis, widespread acantholysis, and focal dyskeratosis (Fig 2). Complete immunobullous workup was negative. Herpes simplex virus testing with tissue polymerase chain reaction was negative. The culture grew *Pseudomonas aeruginosa*.

The patient received a prescription for a short course of oral ciprofloxacin and topical treatments,

Abbreviation used:

HHD: Hailey-Hailey disease

including gentamicin 0.1% ointment, tacrolimus 0.1% ointment, and amitriptyline-ketamine 2%/5% cream. However, the treatments were irritating and ineffective. Subsequently, Castellani paint was recommended for application to affected areas twice daily. The patient recalled that her grandmother, also affected by HHD, used a magenta topical solution in the distant past. She discontinued all other topical and oral therapy and began treatment with Castellani paint alone. Within 3 weeks, she reported complete remission of the skin disease and associated symptoms. The response was sustained overall more than 2 years later. The patient was unable to be examined in person because she did not reside locally; however, clinical improvement was confirmed by evaluation of photographs that she provided.

DISCUSSION

HHD is often refractory to treatment, including topical and oral antibiotics, topical and oral corticosteroids, topical calcineurin inhibitors, and such systemic therapies as cyclosporine, dapsone, methotrexate, and retinoids. Destructive therapies include surgical procedures, laser (eg, ablative, diode, pulsed dye), photodynamic therapy, electron beam radiotherapy, and dermabrasion.³ Recent publications have reported disease improvement with afamelanotide, glycopyrrolate, botulinum toxin, and low-dose naltrexone.^{3,4}

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Fig 1. Clinical image. Macerated and eroded pink plaques affecting the groin.

In 1905, Italian physician Aldo Castellani developed Castellani paint (magenta or carbol fuchsin solution), a mixture of fuchsin, boric acid, phenol, resorcinol, acetone, and alcohol.⁵ Castellani paint has been used in a wide range of clinical applications: chronic hand dermatitis, tinea pedis and cruris, cutaneous candidiasis, and pustular dermatoses of the hands and feet.^{2,6} The paint has local anesthetic, antipruritic, bactericidal, fungicidal, keratolytic, and drying properties. Basic fuchsin imparts a magenta color and has been reported to stimulate granulation tissue formation and reepithelialization.⁶

Castellani paint is available in over-the-counter formulations that are colorless or contain basic fuchsin, which may stain skin and clothing. It can be irritating with initial application; however, irritant reaction improves with time or dilution. Castellani paint is accessible and well tolerated. As a means to target microbes, symptoms, and barrier defects,

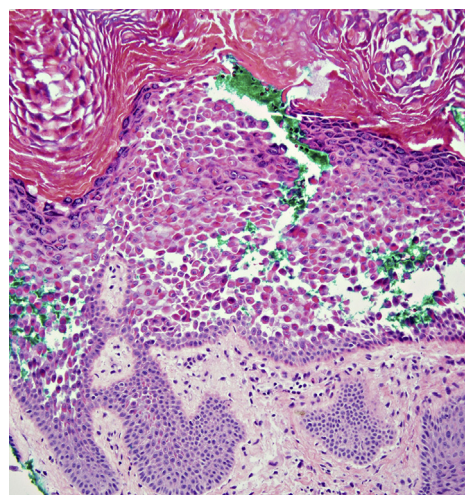


Fig 2. Histopathologic evaluation. Acanthosis, hyperkeratosis, and near full-thickness acantholysis appreciated on histopathologic evaluation (Hematoxylin-eosin stain; original magnification: $\times 100$).

Castellani paint deserves further investigation and consideration as an adjuvant therapy for patients with HHD. An HHD support group recently conducted a survey assessing the use of Castellani paint among its members. Results are being analyzed and prepared for reporting.

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