

Efficacy of topical tetracyclines in Hailey-Hailey disease

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ABSTRACT

Introduction: Familial benign chronic pemphigus or Hailey-Hailey disease (HHD) is a rare autosomal dominant disorder characterized by the development of recurrent blisters and erosions in the intertriginous areas. A variety of systemic and topical treatment options was used with a mixed results, including corticosteroids, topical zinc oxide, topical 5-fluorouracil, topical vitamin D analogs, dapsone, ultraviolet A, systemic retinoids, cyclosporine, methotrexate, and photodynamic therapy, laser therapy and botulinum toxin. **Case Report:** We describe a case of HHD with a history of active hepatitis, which was successfully treated by topical tetracycline after only 2 weeks treatment. **Conclusion:** This report is the first to describe an effect of topical tetracyclines in controlling the skin lesions in HHD.

Keywords: Autosomal dominant disorder, Blisters, Erosions, Familial benign chronic pemphigus, Hailey-Hailey disease, Topical tetracyclines

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INTRODUCTION

Hailey-Hailey disease (HHD, familial benign chronic pemphigus), is an autosomal dominant, hereditary, blistering disorder, characterized by recalcitrant erythema and erosions in the intertriginous area. Although it has been reported that tetracyclines are effective in some blistering diseases [1], the effects of their topical presentation on HHD remain unknown. Here, we report a case of HHD successfully treated with topical tetracyclines.

CASE REPORT

A 35-year-old male with a history of active hepatitis, presented 11 months before his medical consultation a recurrent painful fluid and pus filled lesions followed by painful erosions over the axillae and groins (Figure1). The lesions were mildly pruritic and the discharge was foul-smelling. The lesions aggravated on sun exposure with exacerbation in summer season. He gave family history of similar disease in his father and paternal uncles. Histopathological findings demonstrated acanthosis with suprabasal acantholysis in the epidermis. We diagnosed familial HHD. We prescribed topical tetracyclines twice daily, which promptly improved the skin lesions, within two weeks (Figure 2). Subsequently, the applications were gradually tapered off over a month without recurrence.



Figure 1: Erosions and crusts over the axillae.



Figure 2: Post-treatment photograph after two weeks of starting the topical tetracyclines.

DISCUSSION

Hailey-Hailey disease (HHD) is an autosomal dominant, hereditary, blistering disorder, characterized by recalcitrant erythema and erosions in the intertriginous area. HHD is linked to mutations in the Calcium-transporting ATPase type 2C member 1 gene encoding the human secretory pathway $\text{Ca}^{2+}/\text{Mn}^{2+}$ ATPase. Apart from the symptomatic therapy, drugs such as corticosteroids, dapsone, thalidomide, nicotinamide, etretinate, methotrexate, cyclosporine, topical 5-fluorouracil, topical vitamin D analogs, alefacept, ultraviolet A therapy and photodynamic therapy have been tried with varying results. Laser therapy (carbon dioxide or erbium: YAG laser ablation), botulinum toxin have been reported as useful in treatment of recalcitrant HHD [2–6].

Topical and systemic antibiotic agents in combination with topical glucocorticoids are the mainstay of treatment for HHD [1]. Treatment with antibiotic agents should be initiated if colonization is suspected on the HHD lesions [1]. Although tetracycline is known as an antibiotic, its mechanism of action may include anti-collagenase activity, which may contribute to suppress blistering formation in HHD patients [7]. Collagenase activity depends on the presence of calcium and zinc, which seem to be chelated by tetracycline [7]. Moreover, it is supposed to inhibit neutrophil and eosinophil chemotaxis and lymphocyte transformation, which may also contribute to their anti-inflammatory effect [7]. Topical tetracyclines will reduce considerably the side effects due to systemic treatment mainly because of the chronic nature of HHD.

CONCLUSION

Hailey-Hailey disease (HHD) is a rare genodermatosis that can be challenging for both patients and dermatologists as the disease can significantly impact patient's quality of life and are often difficult to control. To the best of our knowledge, this report is the first to describe an effect of topical tetracyclines in controlling the skin lesions in HHD.

Author Contributions

Amarouch H. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ismaili N. – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Hassam B. – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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