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Single Case

Recurrent Ulcerations in an 84-Year-Old Male Diagnosed with Hailey-Hailey Disease

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Keywords
Hailey-Hailey disease · Autosomal dominant disease · Familial benign chronic pemphigus

Abstract
Hailey-Hailey disease (HHD), or familial benign chronic pemphigus, is a rare inherited acantholytic dermatosis. It is an autosomal dominant disease affecting the intertriginous areas. HHD has been characterized by flaccid blisters, erosions, and macerations that are limited to flexural (friction-prone) areas. The painful blisters and erosions significantly decrease patients' quality of life. There are multiple types of therapy related to this disorder. Many of the studies have suggested benefits from steroid therapy in addition to oral antibiotics.

Introduction

Hailey-Hailey disease (HHD), also known as familial benign chronic pemphigus, is a rare autosomal dominant skin disorder [1, 2]. It is caused by a mutation in the *ATP2C1* gene, which encodes for intracellular calcium pumps that alter the homeostasis of this ion [3–5]. Excessive sweating, heat, and trauma to the skin may exacerbate HHD and cause separation of

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keratinocytes, especially in friction-prone areas [1, 3]. Therapeutic options range from topical to systemic corticosteroids and antibiotics, among others [1].

HHD is not a life-threatening disease, but it may decrease patients' quality of life significantly [6]. We report the case of an 84-year-old male that presented with recurrent nonresolving ulcerations in the mouth.

Case Report

An 84-year-old male was referred to our unit after recurrent oral mucosal erosions involving the oropharynx and buccal mucosa. The patient was known to suffer from Parkinson's disease, atrial fibrillation, hypertension, and dyslipidemia. He presented with a history of chronic oral ulcerations and dysphagia that had not been resolving since the first presentation. He was provisionally diagnosed with recurrent aphthous ulcers and was managed conventionally with oral antiseptics and analgesics.

Incisional biopsy of the buccal mucosa was performed under local anesthesia since the patient's symptoms had not improved with conventional treatment. The differential diagnosis included aphthous ulcers, mucous membrane pemphigoid, erosive lichen planus, and oral squamous cell carcinoma.

Histopathology demonstrated a suprabasal cleft with acantholytic cells and a focal papillaroid projection with dilapidated brick wall appearance (Fig. 1). Also, the subepithelial tissues showed lymphoplasmacytic inflammatory infiltrates (Fig. 2). A suggestive diagnosis of oral HHD was made. Direct immunofluorescence testing is required for confirmation, since it is a pathological diagnosis. Initially, we could not send the patient to direct immunofluorescence testing.

Discussion

HHD is a rare inherited disease that is caused by a mutation of *ATP2C1*, which encodes for a magnesium-dependent calcium pump [6, 7]. It was first described in 1939 by brothers Hailey and Hailey as an autosomal dominant disorder which manifests as recurrent episodes of pruritic vesicles and erosions [4, 8]. It can be exacerbated by sweat, moisture, friction, and ultraviolet radiation [8]. Lesions usually target friction-prone areas such as the axilla, chest, neck, and genital areas [4, 8]. Bacterial infection caused by *Staphylococcus* species is a common feature of HHD [4]. It is of an unknown prevalence [3].

A diagnosis is usually made on histopathological grounds, where suprabasal acantholysis is appreciated. A dilapidated brick wall appearance is often present [3]. The treatments of choice include oral and topical antibiotics, along with systemic and topical corticosteroids [7–9]. Surgical therapies have also been suggested, such as the use of ablation as well as of diode and pulsed dye lasers [9]. In some studies, it has been reported that botulinum toxin injections have ameliorated the skin disease [9].

Statement of Ethics

Written informed consent for publication (including images) has been obtained from the patient. The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Maximum contribution towards the conception, design, analysis of data, and drafting of this manuscript is attributed to the first author, F. Al Qooz. All other authors made equal contributions to (1) the conception and design, as well as the acquisition, analysis and interpretation of data; (2) drafting of the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published.

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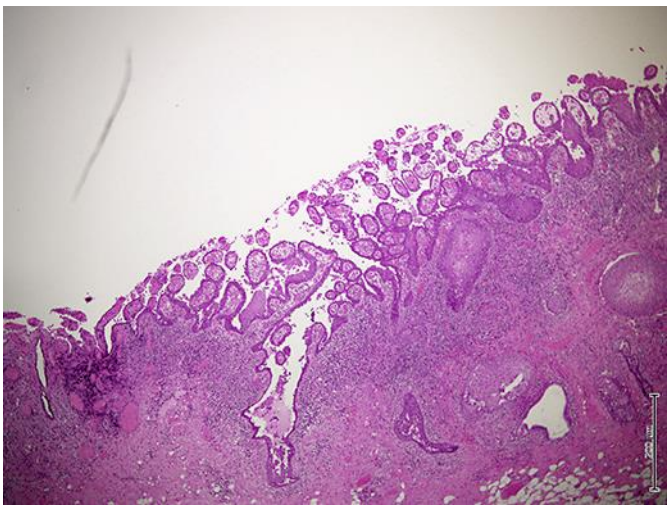

Fig. 1. This section shows suprabasal and intraepidermal clefting with dilapidated brick wall appearance. The subepithelium shows moderate chronic inflammatory cell infiltrate. H&E. ×20.

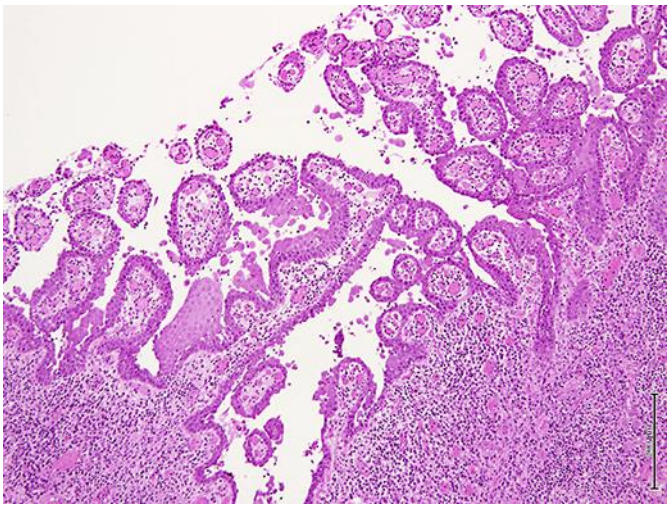

Fig. 2. Closer section showing suprabasal and intraepidermal clefting with dilapidated brick wall appearance. The subepithelium shows moderate chronic inflammatory cell infiltrate. H&E. ×40.