Journal of Pakistan Association of Dermatologists. 2016;**26 (3)**:257-259.

Case Report

Hailey-Hailey disease - therapeutic trial with carbon dioxide laser

Vignesh Karthik Natarajan, Sheela Kuruvila

Department of Dermatology, Venereology and Leprosy, Pondicherry Institute of Medical Sciences, Pondicherry

Abstract Hailey-Hailey disease (HHD; MIM 169600) is a rare chronic autosomal dominant skin disorder resulting from a mutation in APT2C1, which codes for a Ca^{2+}/Mn^{2+} ATPase protein (hSPCA1) in the Golgi apparatus. HHD is clinically characterized by recurrent episodes of vesicles, erosions and erythematous plaques in the intertriginous areas. Triggering factors include trauma, friction, sweating, ultraviolet light exposure, pregnancy, and menstruation. Nail changes include the presence of longitudinal leuconychia. Histopathology shows areas of intraepidermal acantholysis causing the "dilapidated brick-wall" appearance. We present a case of HHD in a 33-year-old male. What was unique in our patient was the painful and therapy-resistant perianal involvement that led to significant morbidity. His mother also had HHD but other family members were unaffected. He had received several treatments earlier including topical antibiotics, antifungals, and corticosteroids without much improvement. We started him on carbon dioxide laser and presently he is under follow-up. We report this case because of its rarity.

Key words
Hailey-Hailey disease, carbon dioxide laser.

Introduction

Hailey-Hailey Disease (HHD: MIM 169600) was first described by Hailey brothers in 1939.[1] HHD is also known as familial benign chronic pemphigus. It is an autosomal dominant disorder with a prevalence of 1 in 50,000 and commonly presents in the third or fourth decade of life. Clinically, HHD is characterized by a recurrent vesicular eruption usually localized to intertriginous regions. The common sites of involvement are neck, axillae, inframammary area, groins and perineum leading to painful fissures. We present a patient with HHD who had disabling perianal involvement and was successfully treated with carbon dioxide laser.

Address for correspondence
Dr. Vignesh Karthik Natarajan
Department of Dermatology, Venereology and Leprosy
Pondicherry Institute of Medical Sciences,
Pondicherry 605014, India
Email: dr.vigneshkarthik23@gmail.com

Case Report

A 33-year-old male presented with complaints of itchy, raised, malodorous lesions over the axillae, external genitalia and perianal area of four years duration. There was history of oral ulcers and he was otherwise medically fit. His mother had similar skin lesions. General, physical and systemic examinations were normal.

There were well-defined erythematous macerated plaques with few vesicles, fissures (rhagades) and erosions seen over the neck (**Figure 1**), axillae, groins, scrotum, root of penis (**Figure 2**), and perianal area (**Figure 3**). Fingernails showed longitudinal leuconychia (**Figure 2**).

Skin biopsy from axilla showed extensive partial loss of intercellular contacts (acantholysis) within epidermis producing a "dilapidated brick wall" appearance (**Figure 4**).

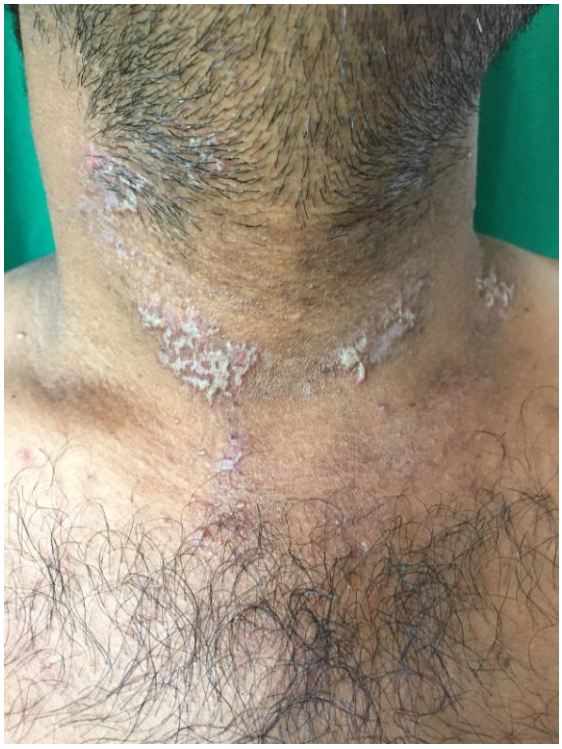

Figure 1 Clinical picture shows multiple vesicopustules on neck.

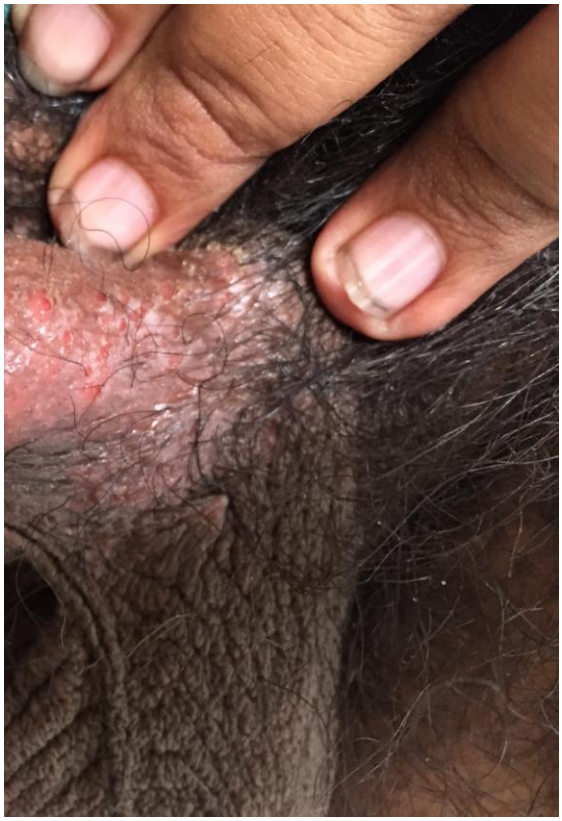

Figure 2 Macerated plaques with fissures on root of penis. Fingernails show longitudinal leuconychia

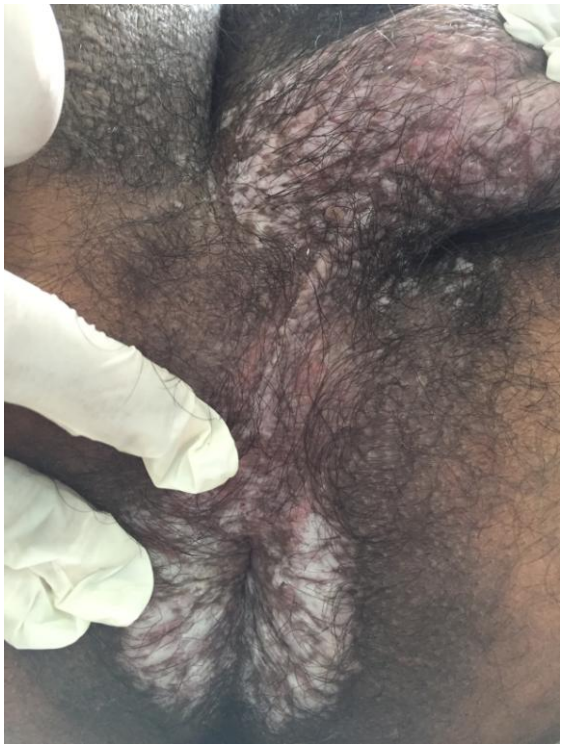

Figure 3 Macerated plaques in perianal area

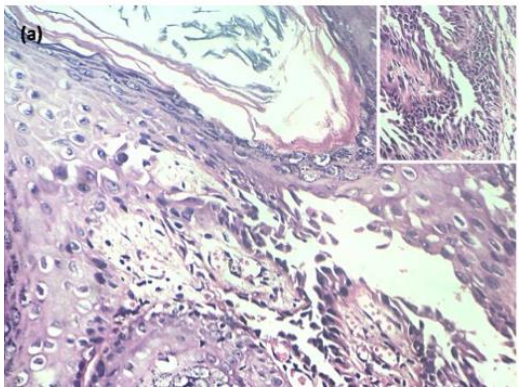

Figure 4 (a) Histopathological examination (H & E, x 40] showing acantholysis producing a dilapidated brick wall appearance (inset, H & E x 40)

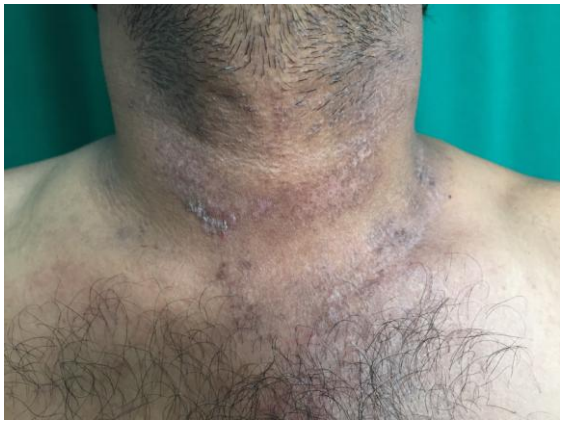

Figure 5 Clinical improvement of the lesions over the neck after two sittings of CO_2 laser

Patient was started on oral doxycycline, topical mupirocin cream and clotrimazole 1% cream. He was also given ablative CO_2 laser (SUDARFRAX^R) once monthly. After 3 months of treatment there was mild improvement in the lesions.

Discussion

HHD is a chronic disease with a significant impact on the patient's quality of life.

In HHD, heterozygous mutation of calcium dependent ATP2C1 gene (chromosome 3q21-24) is seen which results in dysfunction of a Golgi-associated Ca^{2+} ATPase and interferes with intracellular Ca^{2+} signaling.[2]

Morphologically, the lesions have crusted weeping erosions, vesicles, pustules, expanding annular plaques with peripheral scaly borders and vegetating plaques with fissures (rhagades). It is itchy, painful, and malodorous. The mucosae are usually spared.[3] Koebnerization can be present. Longitudinal white bands (longitudinal leuconychia) are seen in fingernails. Clinical course is characterized by exacerbations and remissions.

Histopathologically, there is extensive partial loss of coherence between suprabasal keratinocytes (acantholysis) producing a 'dilapidated brick wall' appearance and clusters of loosely coherent cells float within suprabasal clefts.[4]

Treatment of HHD is difficult. Weight reduction, minimizing friction, control of secondary infections by antibiotics, and judicious use of corticosteroids form the cornerstone of therapy.

Topical treatment includes corticosteroids, antibiotics, retinoids, tacrolimus, calcipotriol and antifungal creams. Corticosteroids, retinoids, dapsone, methotrexate, biologics such as alefacept, cyclosporine and thalidomide are the systemic agents used in the treatment of HHD. Botulinum toxin type A, dermabrasion,[5] erbium:YAG laser, narrowband ultraviolet B treatment and photodynamic therapy have also been tried with variable success. In recalcitrant cases, surgical ablation by CO_2 laser has yielded satisfactory results.[6]

References

1. Hailey H, Hailey H. Familial benign chronic pemphigus. *Arch Dermatol Syphilol*. 1939;**39**:679–85.
2. Sudbrak R, Brown J, Dobson-Stone C, Carter S, Ramser J, White J *et al*. Hailey-Hailey disease is caused by mutations in ATP2C1 encoding a novel Ca2+ pump. *Hum Mol Genet.* 2000;**9**:1131-40.
3. Burge SM. Hailey-Hailey disease: the clinical features, response to treatment and prognosis. *Br J Dermatol.* 1992;**126**:275-82.
4. Thapa DP, Jha AK, Pudasaini S, Kharel C, Shrestha S. Genital Hailey-Hailey disease: A case report. *Our Dermatol Online.* 2013;**4**:87-8.
5. LeBlanc KG Jr, Wharton JB, Sheehan DJ. Refractory Hailey-Hailey disease successfully treated with sandpaper dermabrasion. *Skinmed*. 2011;**9**:263-4.
6. Hunt KM, Jensen JD, Walsh SB, Helms ME, Soong VY, Jacobson ES *et al.* Successful treatment of refractory Hailey-Hailey disease with a 595-nm pulsed dye laser: a series of 7 cases. *J Am Acad Dermatol*. 2015;**72**:735-7.