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Hailey - Hailey Disease - A Case Report

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ABSTRACT

This case report emphasizes the importance of a multidisciplinary approach and provides valuable insights into the clinical course and management strategies for Hailey-Hailey disease. Hailey-Hailey disease is a rare autosomal dominant skin disorder characterized by recurrent blistering and erosions primarily affecting intertriginous areas. We present a case report of a 42-year old male patient with a confirmed diagnosis of Hailey-Hailey disease. The patient had a history of pruritic skin lesions over neck, inframammary areas since the age of 18. The lesions aggravated more during summer than winter. Clinical examination revealed discrete vesicles, erosions, macerations present in the bilateral inframammary areas, flexures and scalp along with crusting. Histopathological examination of skin biopsy taken from right inframammary area revealed supra basal vesiculobullous lesion. Treated with systemic antibacterial, topical antifungals to cover for secondary infection following topical steroids. The skin lesions improved. Then the patient came with complaints of itching & skin lesions on scalp, trunk, groin. At first tab Dapsone 50 mg was started, but the patient was not tolerated and hence it was stopped. Therefore, she was treated with oral antibiotics, topical steroids, antihistamines. Eventually, skin lesions were reducing and improving. Now, in remission and on medications. This case report provides valuable resources of unusual information that may lead to research and advances in clinical practice.

INTRODUCTION

Hailey-Hailey disease, also called Familial Benign Chronic Pemphigus, is a rare autosomal dominant disorder which occurs with remitting and relapsing episodes[1]. It has characteristic features of painful, pruritic, foul smelling vesicles and bullous lesions associated to erosions in intertriginous areas[2]. It occurs due to mutations in the ATP2C1 gene producing a Golgi-associated Ca2+ ATPase dysfunction resulting in suprabasal acantholysis being that trauma, heat and infections play an important role in the exacerbation and persistence of the disease[1].

CASE REPORT

A 60 year old female patient with known history of DM and no history of Thyroid disorder presented to dermatology OPD with history of pruritic skin lesions over neck, inframammary areas since the age of 18. The lesions aggravated more during summer than winter. There was trend of remission and exacerbation. The patient had no family history of similar lesions. On examination, there were discrete vesicles, erosions, macerations present in the bilateral inframammary areas, flexures and scalp along with crusting. No active border for lesion. No pustules. Not foul smelling. No vegetative growth. No thick adherence scaling. She was treated with systemic antibiotics, antifungal, topical steroids. On follow up, macerated erosive lesions presented in groin (FIG 1), inframammary area (FIG 2), neck, scalp. With clinical finding

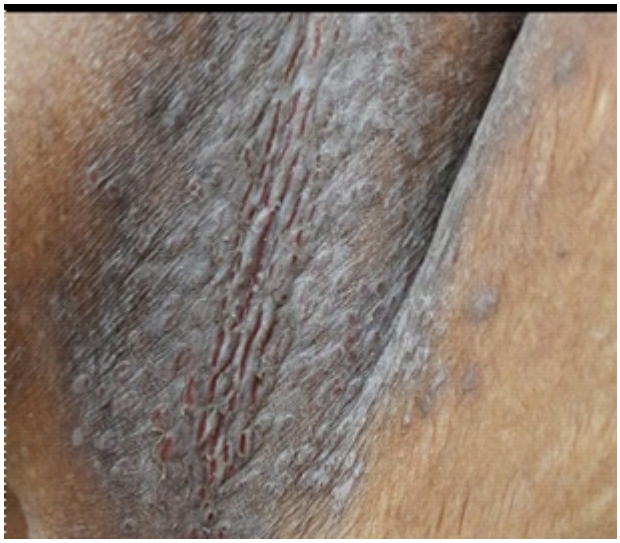

Fig. 1 : Macerated erosive lesions on groin

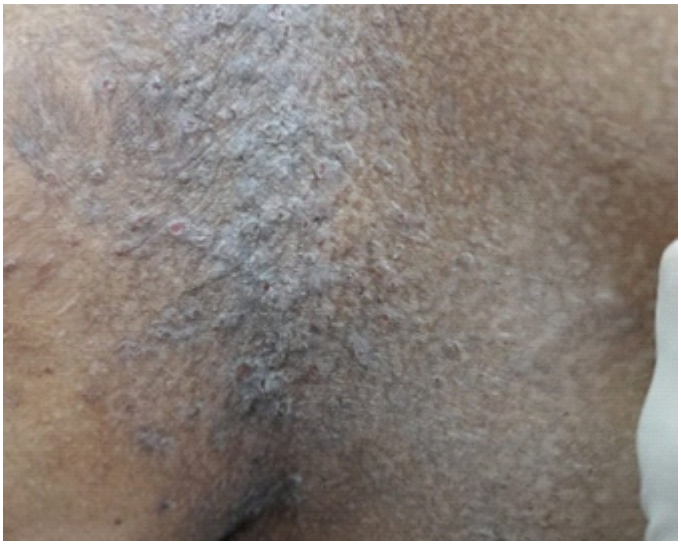

Fig. 2 : Macerated erosive lesions on inframammary area

and chronicity, hailey hailey disease was considered and Skin biopsy taken from right inframammary area revealed supra basal vesiculobullous lesion. Reported as HHD. Treated with systemic antibacterial, topical antifungals to cover for secondary infection following topical steroids. The skin lesions improved and then advised to do Direct immunofluorescence (DIF) study, which was negative. Then the patient came with complaints of itching & skin lesions on scalp, trunk, groin. On examination of flexural, groin, inguinal areas revealed discrete and confluent papules, vesicles, fissury area, maceration and crusted lesions on scalp. So the plan was to start tab dapsone 50 mg alternate days, but the patient had tiredness and disturbance with a single dose and hence it was stopped. Therefore, she was treated with oral antibiotics, topical steroids, antihistamines. Eventually, skin lesions were reducing and improving. Now in remission and on medications.

DISCUSSION

Hailey-Hailey disease is a rare genetic disorder that affects the skin and the incidence of the disease is 1 in 50,000. It is characterized by recurring blisters and erosions in areas of the body where there is friction or sweating such as neck, armpits, groin and under the breasts[3]. Our patient came with recurrent chronic vesico bullous lesions started at the age of 18. It is caused by mutations in the ATP2C1 gene, which provides instructions for making a protein that helps to transport calcium ions across cell membranes[3]. The mutation leads to a deficiency of this protein, which disrupts the normal functioning of skin cells and causes the characteristic blisters and erosions. The condition usually begins in early adulthood and tends to be chronic and progressive, although the severity of symptoms can vary widely between individuals[4]. The differential diagnosis of Hailey Hailey disease includes fungal infection, intertrigo, psoriasis, extra mammary Paget's disease, acanthosis nigricans, pemphigus vegetans and Darier's disease[2].

Its management includes control of exacerbating factors, secondary infections, and cutaneous inflammation[5]. Treatment options include topical and systemic medications as well as surgery in some cases. Topical therapy includes antibiotics, steroids, tacrolimus, and calcitriol, whereas systemic therapy includes antibiotics (clindamycin, gentamicin, and mupirocin), steroids, methotrexate, tacrolimus, dapsone, and thalidomide[5]. Management of our patient includes candid powder, topical steroids like beclomethasone and clotrimazole cream, clobetasol and gentamicin cream, topical antifungal cicloproix olamine, doxycycline as systemic antibiotic, cold saline compressors.

CONCLUSION

The uniqueness of our report is that disease has diagnosed after 19 years of recurrent chronic skin lesions on 2022. After confirming the disease with biopsy she was managed with non-pharmacological and pharmacological measures. Non pharmacological measures include to avoid friction, use loose cotton clothing, weight reduction, condys compresses. Pharmacologic treatment includes systemic antibiotics, topical steroids, candid powder and dapsone.

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CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

ETHICAL STATEMENT

Ethical approval is not applicable for case report in our institution.

REFERENCE

1. Thapa DP, Jha AK, Pudasaini S, Kharel C, Shrestha S. Genital Hailey-Hailey disease: A case report.Our Dermatology Online. 2013;4(1):87.

2. Prateek K, Banwarilal MR, Chaudhary SS, Garg M. Hailey Hailey disease-a rare case report. Int J Res Dermatol. 2016 Apr;2(2):36-9.

3. R. L. Sharma, Rekha Sharma. Hailey Hailey Disease: Case Report of a rare disease. IAIM, 2018; 5(7): 94-98.

4. Sangoram SS, Indurkar VA, Amin-Hon VS, Nishigandh PD. Hailey-Hailey disease-a case report. Pravara Medical Review. 2010;5(4):30-2.

5. Ogbuneke U, Odega E, Ibrahim YW, Abubakar M. A Rare Case of Atypical Recalcitrant Hailey-Hailey Disease and a Literature Review. Case Reports in Clinical Medicine. 2022 Sep 2;11(9):360-5

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