Hailey Hailey Disease: Treated with Magnesium Glycine

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Case Report

ABSTRACT

Hailey hailey disease is an autosomal dominant disorder caused by mutation in gene ATP2C1 leading to loss of cell adhesion and further acantholysis. The disease commonly involves the intertriginous areas and is characterized by erythematous, macerated, malodorous, fissured plaques.Traditional treatment modalities include steroids(topical&oral), retinoids , immunosupressants. We report successful use of magnesium glycine in a chronic non-responding case of HHD.

KEY WORDS: autosomal dominant, benign familial, magnesium glycine, pemphigus

INTRODUCTION:

Hailey -Hailey disease (HHD) is a rare autosomal dominant bullous disorder with prevalence of 1:50000 in a general population, lesions are commonly seen over flexural surfaces in form of flaccid vesicles, bulla, pustules or warty lesions which starts in third or fourth decade. Lesions start with a vesicle or pustule and extend peripherally healing in centre. Watery to pus discharge occurs from the lesion leading to maceration and foul smell affecting quality of life of the patient[1].

CASE REPORT:

A 44 year male reported to out-patient department with 20-years history of recurrent red, moist lesions over intertriginous areas of body over bilateral axilla and groins. The lesions were pruritic and were associated with stinging and burning sensation. Malodor from the lesions caused distress to the patient in professional life. Recurrent episodes of exacerbations of the disease were reported especially in summer. The patient had been treated in the past with both topical and systemic steroids / topical antifungal and antibiotic combination as well as antibiotics but unsatisfactory response lead to discontinuation of treatment. On/off use of systemic steroids for 2 years lead to diabetes mellitus which is the main adverse effect of conventional therapy. Patient's son aged 25 year also started developing similar pustulo-vesicular lesions over neck since 6 months though biopsy was not done but on examination lesions were suggestive with HHD.

On examination, erythematous, macerated, oozy and malodorous plaques with multiple fissures in the axillary, inguinal and perineal areas, (Figure 1). All other cutaneous examination was within normal limits. Patients HBA1C was 8 FBS was 150mg/dLand PPBS was 230mg/dL. All other routine haematological and biochemical investigations were normal. KOH smear for fungus was negative. Tzanck smear showed a few acantholytic cells. A 4 mm punch biopsy specimen was taken from the affected axillary tissue. Intraepidermal blistering dermatitis with numerous incomplete acantholytic cells seen within the blister cavity on biopsy. The epidermis is hyperplastic and shows a suprabasal acantholytic blister at places with tombstone pattern. Incomplete acantholysis extends to the hyperplastic epidermis where it is seen to involve more that half of its thickness (Figure 2). The clinical diagnosis of HHD was thus histopathologically confirmed. Patient was put on oral magnesium glycine 200mg BD daily, topical steroids and Potassium Permagnate soaks over macerated lesion. A significant improvement in the

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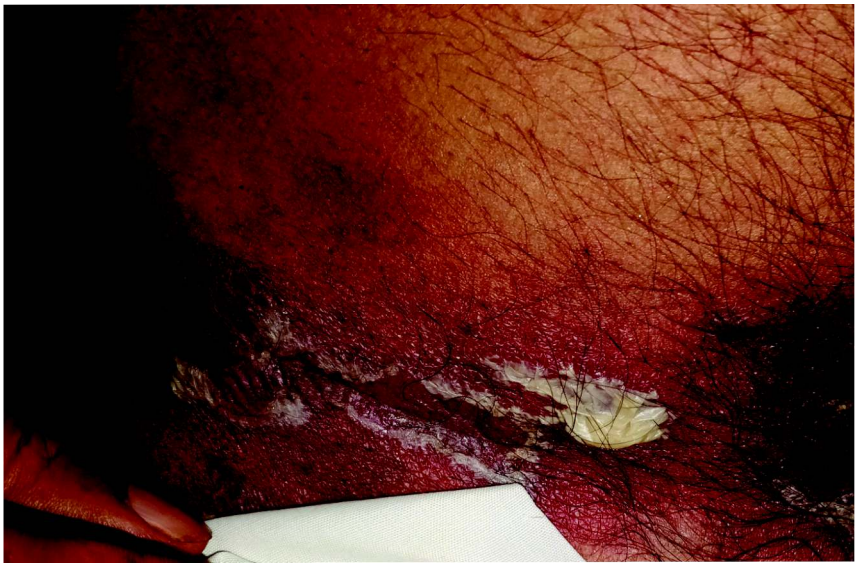

Figure 1: Skin Lesions showing pus filled bulla with maceration in groin.

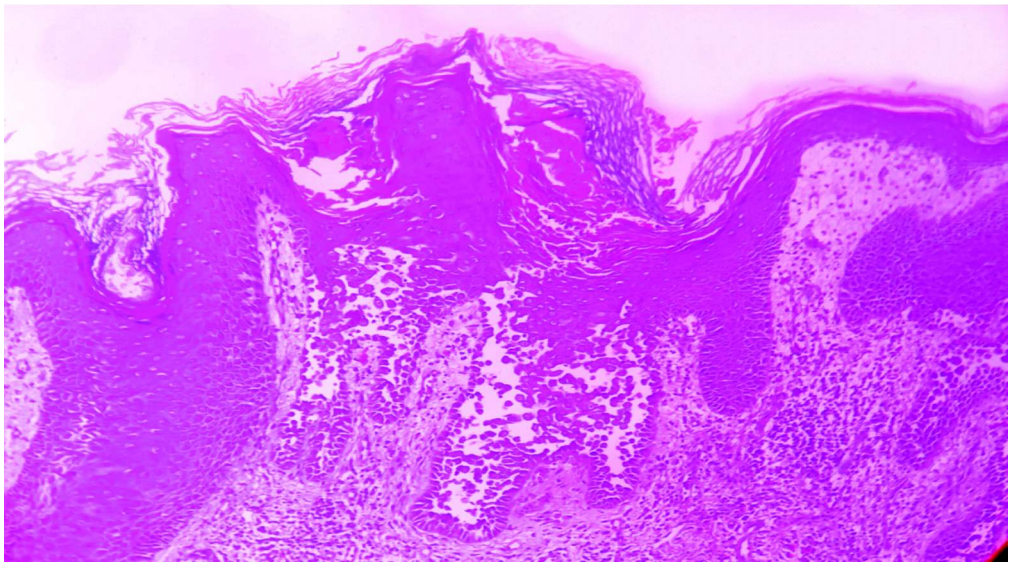

Figure 2: Biopsy shows intraepidermal blistering dermatitis with numerous incomplete acantholytic cells seen within the blister cavity. The epidermis is hyperplastic and shows a suprabasal acantholytic blister at places with tombstone pattern . Incomplete acantholysis extends to the hyperplastic epidermis where it is seen to involve more that half of its thickness (Haemotoxylin&Eosin stain 10x).

skin lesions was noted after 15 days of treatment and an almost complete remission was obtained after 4 weeks as it is a reoccurring disorder drug was planned to be given for 6 months and thantaper (Figure 3).

DISCUSSION:

HHD was first described by the Hailey brothers in 1939.Lesion starts from side of neck and then spreads to axilla and groins. It is aggravated by sunlight, friction, humid season and occlusive clothing. It is caused by mutations in ATP2C1 gene located on chromosome 3q21 that encodes for human secretory-pathway Ca2+/Mn2+-ATPase isoform 1 (hSPCA1) on the Golgi membrane. This gene is expressed by mutated keratinocytes in which they are unable to compensate for partial loss of SPCA1 [2].Normal extracellular Ca2+ concentrations between the basal and superficial epidermal layers is attenuated in HHD with a marked reduction in the total [Ca2+] in the granular layer thus affecting desmosomal components leading to loss of cell adhesion and acantholysis[3-4]. The energy required for

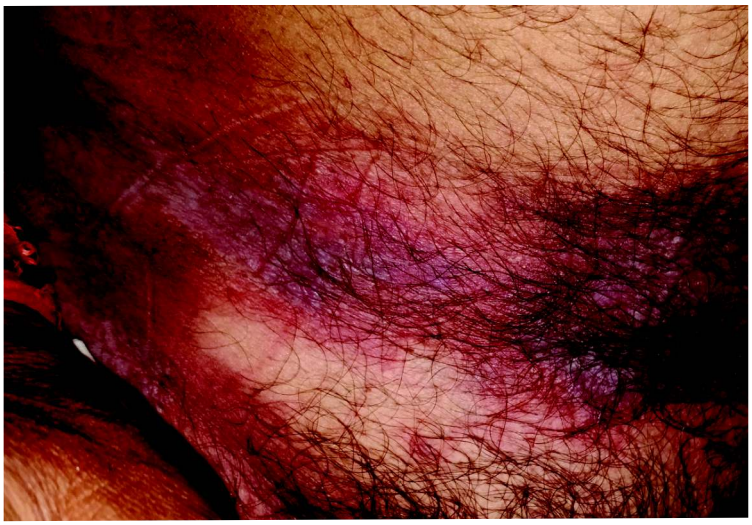

Figure 3: Lesions driedup after 4 weeks of treatment.

all Calcium pumps are derived from adenosine triphosphate by catalyzing hydrolysis in two Magnesium-dependent steps. The Mg^{2+} ion regulates the activity of plasma membrane Ca^{2+}-ATPase. Hence, magnesium supplementation reduces the extrusion rate of Ca^{2+} and increases intracellular Ca^{2+} sequestration. The intracellular calcium to magnesium ratio varies from 4:1 to 1:1. The recommended daily allowance of magnesium is between 360 and 410 mg a day[5-6]. Barde et al used 300 mg magnesium chloride in two cases and reported complete remission in 4 weeks in 1st case and 2 weeks in another[6].

Earlier steroids, immunosupressants, antibiotics and retinoids have been used for the treatment. Many physicians still rely on steroids for the treatment. Relapse is commonly seen in patientsafter stopping the treatment. Outcome of various treatment modalities used in past like cryosurgery, dermabrasion, electrodesiccation have given unsatisfactory results and recurrence is always present.

CONCLUSION:

Treatment with magnesium glycine results in a change in the intracellular calcium hemostasis which may be the molecular basis for the therapeutic response in Hailey–Hailey disease as seen in this case.

REFERENCES

1. Fine JD, Burge SM. Genetic Blistering Diseases.In: Rooks textbook of dermatology .8th Edn.; Vol 2. West Sussex: Blackwell Publishing Ltd; 2010.
2. Callewaert G, Parys JB, De Smedt H et al. Similar Ca(2+)-signaling properties in keratinocytes and in COS-1 cells overexpressing the secretory-pathway Ca(2+)-ATPase SPCA1. Cell Calcium. 2003;34:157–62.
3. Tu CL, Chang W, Bikle DD. The role of the calcium sensing receptor in regulating intracellular calcium handling in human epidermal keratinocytes. J Invest Dermatol. 2007; 127: 1074–83.
4. Mauro TM. CaR talk: the calcium receptor finds new places to park. J Invest Dermatol. 2007; 27: 991–2.
5. Durlach J, Guiet-Bara A, Pagès N, Bac P, Bara M. Magnesium chloride or magnesium sulfate: A genuine question. Magnes Res. 2005;18:187-92.
6. Barde NG, Mishra DB, Ingole SO. Oral magnesium chloride: A novel approach in the management of Hailey–Hailey disease. Indian J Dermatol Venereol Leprol. 2017;83:259-62.

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