



GENETICS AND GENODERMATOSES

SUCCESSFUL TREATMENT OF HAILEY-HAILEY DISEASE WITH ALITRETINOIN

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Background : Hailey-Hailey disease (HHD), also known as familial benign chronic pemphigus, is a rare autosomal-dominant blistering skin disease. HHD has a chronic course and repeats relapse and remission. Bacterial or fungal infections are common. There are various therapeutic options including antibacterial and antifungal agents, corticosteroids, immunosuppressants, vitamin D derivate, retinoides, dapsone, biologics, surgical excision, laser and radiation therapy. But, these treatments are often ineffective or limited.

Observation : We report a case of 50-year old man with 5-year history of erythema, vesicles and erosions with pustular discharge on the axillae and inguinal area. He had been treated with topical corticosteroid, antibiotics and systemic corticosteroids, but there was no improvement of the lesions. A 4-mm punch biopsy was taken from the lesion on axilla. Histology showed intraepidermal clefts, corp ronds in the granular layer and extensive acantholysis throughout the epidermis giving appearance of a 'dilapidated brick wall'. From these findings, we diagnosed the patient Hailey-Hailey Disease (HHD). After being treated with alitretinoin 30mg once a day and topical steroid for 3 months, skin lesions much improved.

Key message : There are various therapeutic options for HHD, but these are often ineffective or limited. Because alitretinoin is well tolerated and has a shorter period of contraception, we suggest alitretinoin as an additional therapeutic option of refractory HHD.

