

Short Communication

Activation of Hailey-Hailey disease by herpes simplex infection in a patient with concomitant two different autoimmune diseases

Sir, our case 61-year-old female was admitted to our dermatology clinic because of severe erythema and pain in her groin. Skin biopsy from her groin and subjected histopathologic examination revealed Hailey-Hailey disease three years ago. She was treated by our dermatology clinic for three years. She had also alopecia universalis for nearly 50 years and Hashimoto thyroiditis for three years. Patient systemic examination was normal. On dermatologic examination, there were sharply demarcated flaccid vesicles, erosions, fissure and crusting lesions in groin (**Figure 1**), axilla, submammary area. She had also alopecia universalis and asymptomatic, longitudinal white bands in the fingernails. Fasting blood glucose, sedimentation, liver function tests, renal function tests in laboratory investigations were within normal limits. Thyroid autoantibody that anti T (>500 U/mL), anti M (>1300 U/mL), free T3(1,83 pg/mL) levels were higher than normal. Thyroid ultrasound was consistent with Hashimoto thyroiditis. Herpes simplex type I and type II IgM were negative and type I and II IgG antibody were negative, positive, respectively.

Depression was diagnosed by psychiatry consultation. Systemic steroids, topical antibiotic and pimecrolimus treatment was started. Unfortunately, symptoms did not improve. So, skin biopsy was taken from patient's groin lesion. Also, herpes simplex lesions were diagnosed histopathologically in the groin (**Figure 2**). Our case had alopecia universalis for 50 years, while Hailey-Hailey



Figure 1 Flaccid vesicles, erosions, fissures and crusting lesions in groin.

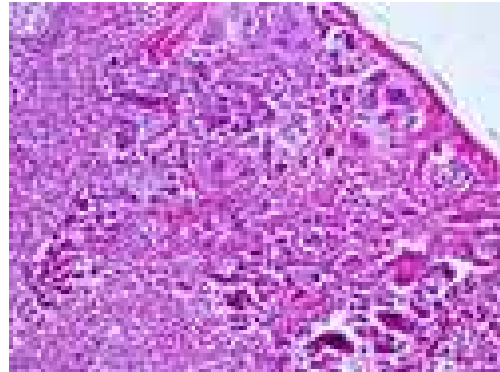


Figure 2 Intraepidermal cleft with multinucleate giant cells.

disease and Hashimoto thyroiditis were diagnosed about three years back. Family history was negative for Hailey-Hailey disease, so we thought that *de novo* mutation.

Discussion

Hailey-Hailey disease is an inherited disease in which intractable, recurrent vesicles affecting the neck, axillae and groins appear. Hailey-Hailey disease is caused in ATP2C1, a gene on chromosome 3q21 that encodes a P-type calcium-transport adenosine triphosphatase (ATPase). *De novo* mutation has been diagnosed in 30% of patients. Hailey-Hailey disease may be triggered by trauma, bacterial, fungal infections, patch test and sunburn.¹⁻³ It is reportedly exacerbated by herpes simplex infection.⁴⁻⁶ Our case with Hailey-Hailey

disease was exacerbated by herpes simplex and regressed by herpes simplex treatment. We want to emphasize the presence of herpes simplex lesion concomitant with treatment-resistant bullous lesion of Hailey-Hailey disease. Hailey-Hailey disease treatment management should bring to mind HSV infections.

References

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Scleredema diabeticorum

Sir, a 48-year-old male presented with spontaneous onset of pigmentation on his upper arms and back followed by development of progressive thickening of the skin which first started from the neck and gradually extended downwards to involve the upper back and arms of 3 years duration. There was no history of infection prior to development of induration. Our patient was a known diabetic since past 20 years on irregular treatment. Cutaneous examination revealed widespread woody hard induration of the skin over the neck and upper back extending upto the arms with sparing of face and hands (**Figure 1**). The skin of the affected areas could not be pinched. Systemic examination was unremarkable.

Apart from a raised blood sugar level, his other hematological and biochemical parameters such as a complete blood count, ESR, liver and renal function tests, thyroid and immunoglobulin profile were normal. Skin biopsy for histopathologic examination showed a thickened reticular dermis with swollen collagen bundles separated by clear fenestrations suggestive of scleredema (**Figures 2 and 3**).

Discussion

Scleredema, a rare collagen disorder, was first reported by Piffard in 1876. The term scleredema is a misnomer because neither sclerosis nor edema is found on microscopic examination. Clinically it is characterized by symmetric diffuse induration of the upper part of the body due to deposition of hyaluronic acid in the dermis.

Graff described three types of scleredema adultorum.¹ Type 1 is the classic type, which was first described by Buschke in 1902. It characteristically develops following a febrile