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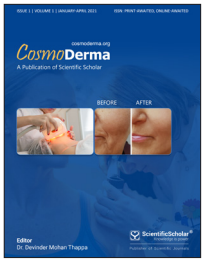

Letter to the Editor

Atypical discoid eczema-like presentation of Hailey-Hailey disease

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Dear Sir,

Hailey-Hailey disease (HHD) is an autosomal dominant inherited acantholytic disorder characterized by a mutation in the ATP2C1 gene.[1] The diagnosis of HHD is based on family history, classical flexural involvement, and clinical morphology of skin lesions. Herewith, we report a case of HHD with atypical discoid eczema-like presentation involving both intertriginous and non-intertriginous areas.

A 22-year-old male presented to the outpatient department with history of vesicles, bullae, and erosions all over the body of 7 years duration associated with remissions and recurrent summer exacerbations. The bullae appeared initially over the neck, upper forearm, and groin which gradually progressed to involve the trunk, bilateral anterior axillary folds, popliteal fossa, and perianal area over the next 20 days. The blisters used to rupture spontaneously over 4–5 days to form oozy macerated plaques with yellow-coloured crusting. The lesions were associated with itching and burning sensation. There was a family history of similar lesions in his mother and grandmother. On examination, he had multiple fissured plaques over the neck, upper back, lower abdomen [Figure 1a], bilateral anterior axillary folds [Figure 1b], upper forearm, lower arm, posterior aspect of left lower thigh [Figure 1c], and groin. Interestingly, there was sparing of the axillary vault, cubital fossa, and popliteal fossa. Palms, soles, mucosa, nails, and hair were not involved. We considered the differential diagnosis of autoimmune intraepidermal blistering disorders, eczema, and HHD. Histopathological examination revealed parakeratosis, suprabasal blisters with acantholytic cells, occasional dyskeratotic cells, and a row of tombstone appearance over the basal layer [Figure 2a and b]. The superficial dermis had perivascular lymphocytic infiltrates. Direct immunofluorescence was negative [Figure 2c]. A final diagnosis of HHD was made based on the clinical and histopathological findings. The patient was started on low-dose oral naltrexone 5 mg once daily and is on follow-up.

HHD (benign familial chronic pemphigus) occurs in 1–4 in 100,000 population. There is no specific gender or ethnic preference. Mutations in the ATP2C1 gene located on chromosome 3q22.1 alter the intracellular calcium gradient in the Golgi complex, resulting in dysfunction of the desmosomal proteins. This leads to acantholysis of the epidermis. HHD presents in the third to fourth decade of life as flaccid vesicles and bullae affecting the axillary, genitocrural, and inframammary folds. These lesions spread peripherally with serpiginous borders to form large, vegetative, malodorous plaques with painful fissures. The flexural disease may be disabling,

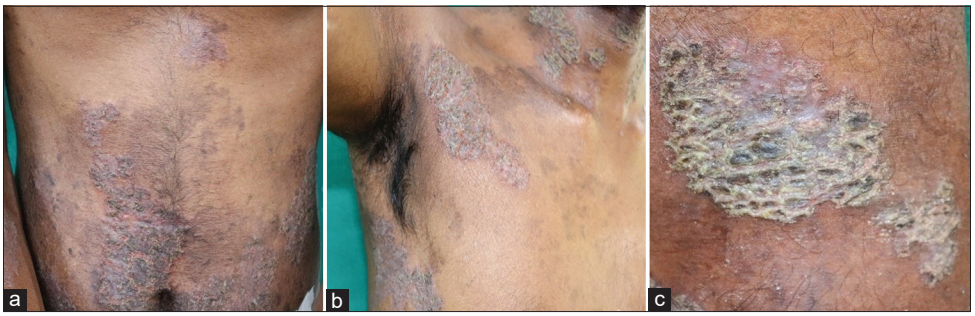

Figure 1: Multiple, well-defined, yellow-colored, crusted plaques present over the (a) lower abdomen, (b) axillary folds, and (c) posterior aspect of thigh.

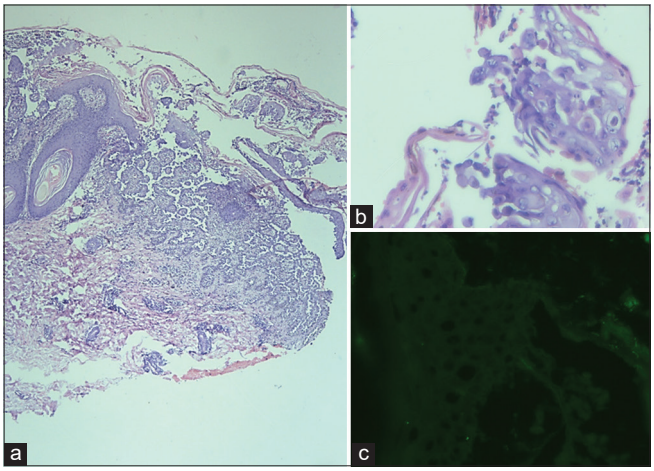

Figure 2: (a) Section shows acanthotic epidermis with suprabasal and intraepidermal acantholysis. Hair follicles and adnexal structures are spared (hematoxylin and eosin stain, ×40). (b) Section shows acantholytic cells with focal dyskeratosis. Parakeratosis and neutrophilic infiltrates in the keratin layer are evident (hematoxylin and eosin stain, ×400). (c) Section shows absence of IgG staining in the epidermis (direct immunofluorescence with immunoglobulin G, DAKO monoclonal antibody, USA, ×400).

especially if it involves the groin and peri-anal area. The presence of vegetation and malodor is clinical indicators of bacterial or fungal infection. Longitudinal white bands in the nails have been described in approximately 70% of patients with HHD. This can be an additional clue to the diagnosis. Mucosal involvement is rare. HHD has a chronic remitting and relapsing course causing significant impairment in the quality of life.[2] Triggering factors for relapse include sweating, friction, ultraviolet radiation, and secondary infection.

Histopathologic examination usually reveals widespread loss of cohesion among suprabasal keratinocytes (acantholysis), resulting in suprabasal clefts. Areas of incomplete acantholysis form layers of intraepidermal and suprabasal detached keratinocytes resulting in a dilapidated brick wall appearance. Dyskeratosis is usually mild. The papillary dermis contains a sparse, perivascular lymphocytic infiltrate with scattered eosinophils. In contrast to autoimmune pemphigus, intercellular immunoglobulin G is absent in the epidermis.

Classical presentation over flexural areas often mimics erosive intertriginous dermatoses such as candidal intertrigo, flexural psoriasis, pemphigus vegetans, seborrheic dermatitis, and Darier's disease. However, lesions over less-occluded areas such as the trunk often show only crusted erosions resembling discoid eczema or annular plaques with peripheral scales.[3] However, in discoid eczema, a spongiotic tissue reaction pattern is seen with massive intercellular edema of keratinocytes and dermis in acute stages. This is followed by hyperkeratosis, acanthosis, mild parakeratosis, and fibrosis of papillary dermis in chronic stages.[4]

Basic measures such as avoidance of ultraviolet radiation, sweat, and friction and wearing loose cotton clothes prevent exacerbation. For patients with milder disease, topical antibiotics and intermittent use of topical corticosteroids or topical calcineurin inhibitors can be used. Patients with severe disease are treated with oral antibiotics, systemic corticosteroids, oral retinoids, and low-dose naltrexone. Surgical and destructive methods have been used in a few patients with recalcitrant disease, including surgical excision, CO_2 laser, 595 nm pulsed dye laser ablation, dermabrasion, and photodynamic therapy.[5] Few case reports have demonstrated favorable response with narrowband UV-B therapy[6] and Vitamin D supplementation.[7]

Our patient had an atypical presentation with discoid eczema-like lesions over the trunk along with involvement of non-intertriginous areas. However, a clinicopathological correlation with a positive family history clinched the diagnosis. This case is reported to highlight the atypical location of skin lesions and difficulties in diagnosing HHD.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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