Hailey–Hailey disease

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DISCUSSION

The prevalence of Hailey–Hailey disease (HHD) also known as familial benign chronic pemphigus is 1:50,000.[1] It is an autosomal dominant disorder caused due to mutation in ATP2C1 gene.[2,3] The gene encodes an adenosine triphosphate–powered calcium pump in the Golgi apparatus of epidermal cells.[4] Faulty calcium pump action leads to disorganized function of desmogleins, which are calcium-dependent adherence proteins (cadherins). Flaccid vesicles or bullae are the primary lesions in Hailey–Hailey disease. These lesions rupture easily leaving behind macerated erosions. Most commonly involved sites are the neck, axillae, and groins. Sometimes lesions can also appear on scalp, antecubital or popliteal fossa, and trunk. Conjunctiva, mucosa, and vulva involvement is rarely seen. A positive family history of HHD is present in many patients.[5] The management of HHD is challenging. At present, there is no reported cure for HHD. The treatment is primarily aimed at symptomatic relief. A concoction of topical antibiotics; antifungal agents; as well as systemic, topical, and intralesional corticosteroids have been found to be useful in the management of HHD in many cases.[5,6] Other drugs that have also proved to be effective are cyclosporine, retinoids, botulinum toxin A, and dapsone.[1] Recalcitrant plaques in HHD give better result with ablative lasers such as carbon dioxide lasers and erbium:YAG laser.[7]

Figure 1: Photographs showing macerated hyperpigmented plaques over the groins

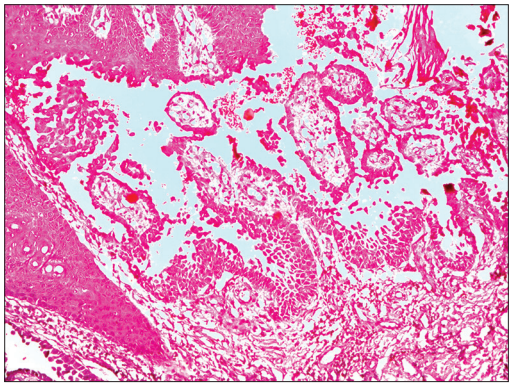

Figure 3: Histopathology slide showing suprabasal clefting with a few acantholytic cells and dilapidated brick wall appearance H and E, X10

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Nil.

Figure 2: Photographs showing macerated hyperpigmented plaques over the neck

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Conflicts of interest

There are no conflicts of interest.

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