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Short Communication

HAILEY-HAILEY DISEASE: AN AUTOSOMAL GENETIC DISORDER

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A B S T R A C T

Hailey-Hailey disease (HHD), also termed as familial benign chronic pemphigus, is a rare autosomal dominant blistering skin disease with waxing and waning in its clinical course. It is described by the presence of flaccid vesiculo-pustules, crusted erosions or expanding plaques in the areas of friction such as neck, axilla, groins, and perineum. Histopathologically exhibits suprabasal separations, inconspicuous dyskeratosis, acantholytic cells within the epidermis, giving a dilapidated brick wall appearance. This article impart about the clinical and histopathological features of HHD and various differential diagnosis for this disease.

INTRODUCTION

Hailey-Hailey Disease (HHD) is an uncommon autosomal dominant Genodermatosis characterized by the development of flexural erosions, blisters, and warty papules.[1] It is also termed as Familial benign Pemphigus or familial benign chronic Pemphigus was originally described by Hailey brothers in 1993. Approximately two-thirds of patients have a family history of the disorder. A history of various relapses and remissions is typical. There are so many hypotheses exist concerning the pathogenesis of familial benign Pemphigus. [2]
It is considered to consequence from a genetic defect in a calcium pump protein. The pump mutation is in *ATP2C1*, a gene bounded on chromosome.[3] The genetic defect in Darier disease, which is a calcium pump defect, is also similar to this gene defect. In addition to the primary gene defect in HHD, contributing factors like heat, friction, and infection are known to exacerbate the disease.[3]

The disease first manifests itself during adolescence or young adult life, although there are occasional exceptions. There is no apparent predilection for occurrence in either gender. The lesions themselves develop as small groups of vesicles appearing on normal or erythematous skin, which soon rupture to leave eroded, crusted areas. These lesions then appear to enlarge peripherally but heal in the center.

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Nikolsky's sign may be present.[4] Nikolsky's sign is a clinical dermatological sign, named after Russian physician Pyotr Nikolsky (1858–1940). (The sign is formed after slight rubbing of the skin results in exfoliation of the outermost layer). It has been frequently noted that heat and sweating simplify the outbreak of the lesions while spontaneous remissions may occur in cold weather. The lesions themselves develop most commonly on those areas of skin which are exposed to friction, e.g. flexure surfaces of the axillae, groin, the neck and the genital area. Tender and enlarged regional lymph nodes may also be present.[1]

It has been recognized that bacterial infection also appears to precipitate the appearance of lesions, and more recently, infection by Candida albicans has been implicated.[3]

Oral lesions occasionally occur in patients with familial benign chronic Pemphigus and mimicking to those occurring on the skin. The lesions develop as crops of vesicles which rapidly rupture leaving raw corroded areas.[1]

The histological appearance of the epithelial lesions in familial benign chronic Pemphigus bears remarkable similarity to that seen in Pemphigus vulgaris and in keratosis follicularis or Darier's disease. Histopathological examination from the biopsy specimen revealed hyperkeratosis and acanthosis with a subcorneal bulla showing intraepidermal clefting and acantholytic cells giving a dilapidated brick wall appearance, which was consistent with the diagnosis of Hailey Hailey disease (HHD). Although the classic presentation and nail findings made the diagnosis straightforward in few cases,

dermoscopy can be a useful clue in the diagnosis of HHD when faced with an intertriginous rash.[1] Other differentials of HHD include sub corneal pustular dermatoses (SCPD), Pustular psoriasis, Intertrigo, Pemphigus vulgaris, Darier's disease, Fungal infections, Acanthosis nigricans, Pemphigus vegetans and Galli Galli disease.[1,3] More studies are required in this area to delineate the dermoscopic differences of these disorders.[1]

Currently an effective result of Minocycline hydrochloride has been reported in a case of HHD.[5] Familial benign Pemphigus waxes and wanes in intensity. Soothing compresses (Aluminum acetate) succeed by intermittent application of topical antibiotics (Clindamycin or erythromycin) and mild corticosteroid preparations results in transient improvement. Erythromycin, Tetracycline and Methotrexate are favored. Bacterial culture and sensitivity may assist guide appropriate therapy. [3]

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