A CASE SERIES OF HAILEY HAILEY DISEASE IN MALE AND FEMALE PATIENTS

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ABSTRACT:

Hailey-Hailey disease was originally described by the Hailey brothers [Hugh Edward and William Howard] in 1939. It is a genetic disorder that causes blisters to from on the skin. The other synonyms of Hailey-Hailey is familial benign chronic, pemphigus. HHD is characterized by out breaks of rashes and blisters on the skin. Affected area of skin undergoes ruptured blistering and inflammation and may be painful to the touch. Area where the skin folds as well as armpits, groin, neck, buttocks and under the breasts are most commonly affected. Here, we discussed about the clinical and various differential diagnosis and treatment of HHD.

Key words: Hailey-Hailey disease, familial benign chronic pemphigus, skin disease.

INTRODUCTION:

Hailey-Hailey disease [HHD] also known as familial benign chronic pemphigus it is a rare blistering skin disease first described by an Howard and Hugh Hailey in 1939.[1] Erythema, Papulovesicular and blisters developed especially where skin folds and other regions exposed to increased friction such as retroarticular region, lateral parts of the neck, axillae, flex or sides of the elbows, navel, inguinal region, genital and perianal region in women and also in sub mammary region.fig-1[2] .It caused by heterozygous mutations in the **ATP2C1** gene that encodes the secretory pathway Ca^{+2}/Mn^{2+} ATP-ase Protein of the Golgi apparatus, leading to alteration in Ca^{2+} - dependent intracellular signaling and resulting in the loss of cellular adhesion in the epidermis.[2]The clinical diagnosis is made based on monomorphic eruption of dome-shaped blisters and pustules. in the eczematous lesions combined with sereve systemic disease, but atypical variations with widespread slits can also happen. The pathogenetic role of topical corticosteroid use in EH is hypothesized, However the risk factor for EH are mainly understood. Objectives and procedures The clinical characteristics and risk factors for EH were characterized. Retrospective research was done on 105AD control patients and 100 EH patients seen between 1980 and 1996.[3]

Hailey-Hailey disease and Eczema herpeticum do not frequently co-exist.The medical literature in both English and german has documented similar cases.[4]

CASE-1

A 40 years old women presented to dermatology outpatient department with history of erosions over the chest and buttock, behind the ear and scalp since 3months associated with itching. Patients was apparently asymptamatic one month ago after which she developed tiny raised lesions over the viginal area associated with itching which gradually increased to form erosions associated with exudation. She developed similar lesions over the chest and breast. There is no family history found. There is past history of hyperthyroidism on regular medication of Tab. Thyronorm 100mcg.There is positive past history of similar complaints in the past since 20years which resolved on topical medications (steroids) and oral steroids and antibiotics. On clinical cutaneous examination diffuse maceration with erythema and yellow coloured exerdat is present over the mons pubis, labinmajura, inguinal folds, extending on the thighs and abdomen. Diffuse maceration positive over the centre of the chest on the intramammary folds with peripheral scaling. Maceration is positive between the buttocks. Moreover on scalp examination multiple scaly plaques positive over the scalp 2*2 involving the hair line nails, oral and genital mucosa and hair is normal. Lab investigations including a complete hemogram, liver and renal function tests within normal limits. Skin biopsy was taken from the lesion and sent for pathological examination. Histopathological examination showed epidermal hyperplasia, suprabasilal cliffing inconspicuous dyskrotosis and acantholysis of keratinocytes resembling dilapidated brick wall .These microscopic features confirmed the clinical diagnosis of Haiely-Haiely diseases.’

CASE-2

A 35-year-old male with a history of erosions over the buttock, behind the ear, and on the scalp since four months along with itching presented to the dermatology outpatients department. Apparently asymptotic for 15days, the patient then developed slightly raised lesions over this ear area that were itchy and slowly grew to become degradation with exudate. similar tumors appeared on chest. No family history was discovered. On a regular dose of Tab .Metformin 500mg there is history of diabetes Miletus. Similar problems have been successfully treated with topical medicines (steroids), oral steroids, and antibiotics over the past 20 years. Upon clinical inspection of the skin, there is diffuse maceration with erythema and yellow-colored exerdat across extending on the thighs and abdomen, with peripheral scaling. Positive maceration occurs between the buttocks. Additionally, normal hair, nails oral, and scalp. Lab tests comprising a full hemogram, liver function tests, and renal function tests that were within normal ranges. From the lesions, a skin biopsy was obtained and sent for pathological analysis. Histopathological analysis revealed acantholysis of keratinocytes that resembled a crumbling brick wall, supraspinal cliffing subtle dyskeratosis, and epidermal hyperplasia. The clinical diagnosis of Haiely-Haiely illness was supported by these microscopic characteristics.

DISCUSSION

The condition is clinically patients presents with a long history of wax and wanes of symptoms, these include as recurrent vesicles, erosions, and maceration in flexural area frication areas , mainly in the axillae , sub mammary folds, groin perineum and neck.[5] In our case the lesions over the viginal area associated with itching and burning sensation. The clinical course or treatment of these disease is difficult to predict , but most of the patient having symptoms of waxing and waning severity.[6]

The secondary viral, fungal bacterial microorganisms are known to associated with Haiely-Haiely disease and the cell carcinoma may associated with Haiely-Haiely.[7] The histophathological criteria diagnosed as Haiely-Haiely with an epidermal hyperplasia, suprabasial clefting inconspicuous dyscrotosis and acantholysis of keratinocytes: due to between index of Hailey-Hailey disease the diagnosis may be misdiagnosed sometimes. Many times, it may mistake with the fungal and bacterial disease.[8] The Dariers disease also have a same clinical features with Hailey-Hailey disease. However, they have different pattern of distribution V-shaped notches of edge of nails plates and palmar pits maybe present in darers disease.[9]

The condition is clinically treated by the first line agents for treatment includes topical antifungals and oral antibiotics.[10] In this patients were treated by steroids, antifungals and antibacterial drugs which is reported as effective.

CONCLUSION

Because Hailey-Hailey disease is a rare disease with a chronic nature, sporadic spontaneous remissions, many recurrences treatment should b personalized. Our patient's response to azathioprine was so positive. That may consider any potential side effects.

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