A rare case: Hailey-Hailey disease

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Abstract

The Hailey-Hailey disease is a rare autosomal dominant disorder triggered by a genetic mutation that is characterized by the growth of repeated blisters and erosions in the intertriginous areas. Commonly the treatment is focused on the symptoms and severity of the condition. Currently, there is no specific treatment available for this disorder.

Keywords: Autosomal disorder, Gene Mutation, Hailey-Hailey disease

Introduction

Hailey-Hailey disease (HHD) is a rare disorder. Due to the lack of accurate diagnosis and many patients not seeking treatment, there is no data available on the prevalence rate. Hailey-Hailey disease is a genetic skin disease induced by mutations in the ATP2C1 gene. This disease is hereditary in an autosomal dominant manner. The signs and symptoms of this rare disorder include blisters and sore rash in skin folds such as the neck, armpits, under the breasts, groin, and in between the buttocks. Symptoms are seen in the months of summer due to heat, sweating (excessive perspiration), and friction (NORD, 2017). HHD is difficult to treat, even with combinations of topical and systemic antibiotic and immunomodulatory medications (Fernandez, 2017). Thus, HHD causes significant morbidity and adverse effects on quality of life (QOL) for affected patients.

Case report

During a home visit, a 53 years old married woman had complaints of persistent itchy, recurring painful lesions filled with pus on the abdomen, for 10 years. The lesions were oozing with foul-smelling discharge most of the time. These lesions intensified on exposure to the sun during the summer. The first episode was ten years back when she suddenly noticed the appearance of 4 – 5 itchy vesicles and also suffered from pain and watery discharge. After two days, the number of vesicles increased to 8-10. The woman consulted a local doctor after two weeks and for which she was prescribed anti-allergens and local application. It healed within 2 – 4 days with crusting and hyperpigmentation. She was temporarily relieved of her symptoms, yet similar lesions started developing on her abdomen, back, armpit, neck, medial aspect of the thigh, and groin, and she has taken treatment for the same. The woman reports saying that her father had the same problem, but no history of fever.

Later on, she has observed the increase in the size of the vesicles to bulla and burst forming, and shallow ulcers with subsequent lesions. The episodes of lesion occurrence gradually increased every second month. She consulted a dermatologist in tertiary care hospital and prescribed Tab Bactrim DS (BD) Tab Clotrimazole 800mg (BD), Tab Hifinac (SOS), and Clonate F cream (local application). The lesions were completely healed, and there was a significant improvement in the lesions. Symptoms were reduced temporarily with the treatment but were not cured; instead, she had episodes of similar legions occurring on her body on and off. The woman tried many oral and topical antibiotics, corticosteroids, which resulted in partial or temporary improvement.

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