AUTOIMMUNE BULLOUS DISEASES

ATYPICAL CASE REPORT OF HAILEY-HAILEY'S DISEASE

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Background: Hailey-Hailey's disease, also called Familial Benign Chronic Pemphigus, is an uncommon autosomal dominant disorder characterized by recurrent blisters and erosive and crusty lesions in intertriginous areas, which resolve without scarring and usually appear more commonly between the third and fourth decade of life. It occurs due to mutations in the ATP2C1 gene producing a Golgi-associated Ca2+ ATPase dysfunction resulting in suprabasal acantholysis being that trauma, heat and infections play an important role in the exacerbation and persistence of the disease.

Observation: A 36-year-old Caucasian woman from the city of Rio de Janeiro, Brazil, complaining of bilateral and symmetrical infra-mammary ulcers and crusted lesions in outbreaks 10 years ago, progressing for 5 months with generalized pruritus with erythematous areas and blisters that rupture presenting crusts and erosions in the infra-mammary areas, thighs, upper limbs and mainly posterior trunk, sparing armpits and groin. Patient reported having a mother with a genetic skin disease but did not remember what it was. A skin fragment biopsy was performed with histopathological findings of Hailey-Hailey's Disease. Patient was treated with vaseline jelly, cephalexin 500 mg orally every six hours for seven days and prednisone 40 mg oral per day for three weeks showing improvement of the lesions.

Key Message: Hailey-Hailey's disease usually prefers intertriginous areas such as folds, armpits and groin, and this case report shows a patient with lesions in atypical areas of the body, causing discomfort and anguish to her for not having a certain diagnosis, showing that the skin biopsy was fundamental added to the family history and the clinical characteristics to make the diagnosis and then, to be able to prevent new complications and to bring greater physical and emotional comfort to the patient.