



GENETICS AND GENODERMATOSES

HAILEY-HAILEY DISEASE: A NEW THERAPEUTIC APPROACH

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Background: Hailey-Hailey Disease (DHH) or Familial Benign Chronic Pemphigus is a rare, autosomal dominant genodermatosis, triggered by a change in cadherin processing, calcium dependent proteins responsible for adhesion between keratinocytes, leading to spontaneous acantholysis or secondary to friction. There are several proposed treatments, including the use of corticosteroids, immunomodulators, sulfones and even surgical therapy with dermabrasion in refractory cases. The aim of this report was to demonstrate that doxycycline is a therapeutic option with favorable results, good tolerance and low rate of side effects.

Observation: Woman, 54 years old, with history of lesions exulcerated in armpits, inframammary region, inguinal and perianal for 6 years. In corticoterapia since then with periods of improvement and worsening. Due to the prolonged use of corticosteroids and their side effects (weight gain, stretch marks, osteoporosis), 100 mg doxycycline was introduced every 12 hours and weaned from corticosteroids. It evolved with healing of the lesions after 30 days. Made gradually reducing the dose, currently maintaining stability with 50mg of doxycycline.

Key Message: DHH is characterized by the appearance of lesions between the 2nd and 3rd decades of life, with a positive family history in 2/3 of the cases. It presents as erythematous plaques with vesicles and flaccid blisters and erosions in the intertriginous areas, and there may be pruritus, burning and local burning sensation. Histopathologically there is an intra-epidermal, acantholytic, supra-basal bubble called the "dilapidated brick wall". Systemic therapy is indicated in more severe cases. Antibiotics of the tetracycline class have anti-inflammatory, antibiotic and immunomodulatory effects. Doxycycline has an average half-life of 20 hours. Its most commonly reported side effect is gastrointestinal intolerance. It presents good absorption, low cost and low toxicity, and can be administered orally, proving to be effective in the reviewed works, being a good option to replace the prolonged corticoterapia

