INTRODUCTION

Familial chronic benign pemphigus is also known as Hailey–Hailey disease. This rare chronic genetic skin disease leads to the formation of vesicles and erythematous plaques with overlying crusts in the axillary and inguinal regions. [1] Treatment of the disease is usually done with antibiotics and topical or systemic corticosteroids although both have limited results, it is highly unusual for cases to present so much resistance to the standard treatment, With the patient taking treatment over 20 years in the past without success, Treatment with botulinum toxin type A is proposed due to its action by reducing sweat production. [2-4]

CASE REPORT

A 60-year-old Caucasian woman with a suspected diagnosis of Hailey–Hailey disease since 20 years, presented with recurrent lesions in the inguinal region, which has made use of different conventional drug treatments for 20 years without results. Treatment with application of botulinum toxin type A showed a significant improvement after about 20 days. The satisfactory response has been linked to decreased local sweating caused by botulinum toxin, which would lead to a less local irritation factors and lower colonization of microorganisms.

DISCUSSION

Hailey–Hailey disease is benign, but often undermines the physical and emotional well-being of patients. Pain, itching, unpleasant odor, and even the appearance of the lesions compromise the quality of life. For this reason, treatment even for cases with few lesions is recommended. [2] The disease usually starts between the second and third decades of life, evolves in spurts, has variable manifestations and the lesions heal without the
formation of scar tissue. There is a mutation in the gene located on chromosome ATP2C1 3q21q24, leading to loss of cell adhesion in epidermis. Epidermal defect leads to acantholysis, either spontaneously or as a result of friction, heat, sweat, or infections. It manifests itself with papules, vesicles, and bullae that break down quickly, leaving erosions and crusts with or without itching or burning, often the lesions present are infected by bacteria, yeasts, and viruses, manifesting usually by fetid odor, pain, and Oozing. The infected tissue plays an important role in the exacerbation and persistence of the disease. The most affected regions by these lesions are the intertriginous areas such as the axillary folds, the inguinal and perineal region, and the sides of the Neck. Mucosal involvement is rare. Histologically, the epidermis shows intraepidermal suprabasal acantholysis and pronounced dyskeratosis of keratinocytes with grains and round bodies. In more chronic lesions, epidermal hyperplasia, parakeratosis, and focal crusts can be observed. Direct immunofluorescence of the lesions is negative. Treatment is symptomatic with general measures to prevent friction and Sweat. For the treatment of secondary bacterial colonization use of the following drugs has been suggested: or al tetracycline, imidazole, sulfone, thalidomide, prednisone, methotrexate and topically cyclosporine, tacrolimus and topical corticosteroids are the most used. Surgical treatment with wide excision, dermabrasion, erbium (yttrium aluminum garnet) or CO₂ lasers, and photodynamic therapy has also been suggested as possible treatments with varying degrees of success. Botulinum toxin type A is a protein that produces chemical denervation, by blocking the release of acetylcholine from nerve endings. The proposed use of botulinum toxin is due to its action blocking cholinergic stimulation by the postganglionic sympathetic fibers, decreasing sweat production by the sweat glands. Heat and sweat are exacerbating factors in patients with Hailey-Hailey Disease. Less sweating, with a reduction of local humidity, works to reduce local irritation factors important in the formation of the lesions and reduces colonization by microorganisms involved in exacerbations. The use of botulinum toxin type A can be considered a safe therapeutic option with simple application in patients with limited response or intolerant to conventional treatments.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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