Answer to Dermacase *continued from* **page 1157**

3. Benign familial chronic pemphigus

First described in 1939, benign familial chronic pemphigus (also known as *Hailey-Hailey disease*) is a chronic, autosomal dominant, cutaneous disorder commonly affecting adults in the third and fourth decades of life.[1,2] Clinically, it is characterized by recurrent, pruritic, painful, and malodorous patches or plaques and vesicular or crusted lesions that mainly affect intertriginous areas such as the axillae and groin, as well as the neck and back.[1,2] This erythematous eruption is commonly exacerbated by hot weather, sweating, stress, friction, and infection. Secondary bacterial and fungal infections are common. Affected patients typically have a history of relapses and spontaneous remissions, but without any residual scarring.[1,2] Results of histology usually reveal rare dyskeratotic cells and widespread intraepidermal acantholysis (loss of intercellular bridges between epidermal keratinocytes), which give the epidermis a dilapidated "brick wall" appearance.[1-3] Results of direct immunofluorescence are usually negative. A biopsy of the lesions in the groin area in our patient had similar characteristic findings.

Genetically, benign familial chronic pemphigus results from mutations in the adenosine triphosphatase–2C1 gene, whose protein product is present in the Golgi apparatus and is involved in regulating intracellular calcium levels.[4,5] Loss of expression of this protein results in calcium depletion from within the Golgi apparatus, which might in turn impair complete processing of junctional proteins that are required for normal cell-to-cell adhesion.

Differential diagnosis

Clinical differential diagnosis of benign familial chronic pemphigus includes tinea cruris, hidradenitis suppurativa, and intertrigo.[6,7] Tinea cruris is a superficial fungal infection of the groin that presents as pruritic, well-demarcated, scaly, and erythematous patches.[6] Diagnosis can be easily confirmed by potassium hydroxide smear tests. Hidradenitis suppurativa is a chronic inflammatory disorder that typically affects apocrine gland regions, such as the axillae, periareolar area, and groin.[7] Patients present with recurrent papules and nodules, which can lead to the development of sterile abscesses, fistulas, and sinus tracts. Intertrigo is a superficial inflammatory dermatitis that commonly involves the skin folds (areas prone to continuous friction) of obese people.[6] It usually presents as erythematous patches with erosions, fissures, and exudate; the eruption is typically exacerbated by hot and humid weather and might succumb to secondary infection.[6]

Management

There is no definitive treatment or cure for this condition. In many patients, however, topical corticosteroids are an effective treatment measure, especially when combined with topical antimicrobials and cleansers.[2,3] Intermittent corticosteroid use with the lowest effective potency is recommended to minimize potential side effects, such as atrophy and striae. In refractory cases, other treatments, including topical creams (tacrolimus, 5-fluorouracil), surgical intervention (excision, laser ablation, photodynamic therapy), and systemic medications (oral antibiotics, corticosteroids, cyclosporine, oral retinoids), might be beneficial.[2,3]

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Competing interests
None declared

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