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Case Letter

Correcting a 30-year diagnosis: A report of vesiculobullous Darier disease previously diagnosed as pemphigus vulgaris

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Dear Editors,

A 55-year-old woman presented with a flare of a blistering rash that began 30 years ago, which was previously diagnosed as pemphigus vulgaris (PV). She endorsed a strong maternal family history of a similar rash and in three of her six siblings. All family members were given the diagnosis of PV. Her blisters flared with stress and illness and previously responded to topical and systemic steroids. The patient endorsed multiple previous biopsies; however, no outside records were successfully obtained due to poor recollection of where the biopsies were performed.

On physical examination, the patient had several flaccid bullae with erythema distributed on the dorsal feet and digits. Additionally, erythematous scaly papules and plaques were appreciated on her inframammary folds (Fig. 1), axilla, medial thigh, and legs. There was no oral, ocular, or genital involvement, but she reported having involvement historically. V-shaped notches were noted on the nails. Punch biopsy was performed of the right distal shin and processed with hematoxylin and eosin (Fig. 2) and direct immunofluorescence. Given the patient's reported history of PV, she was started on intravenous treatment with solumedrol 60 mg daily, doxycycline 100 mg orally twice daily, and niacinamide 500 mg orally three times daily while biopsy test results were pending.

The hematoxylin and eosin test demonstrated acantholysis of the epidermis with dyskeratosis of the keratinocytes in the form of corp ronds and grains. Direct immunofluorescence test results were negative. Testing revealed normal levels of desmoglein-1 immunoglobulin (Ig) G, desmoglein-3 IgG, and bullous pemphigoid 180 and 230 IgG antibodies by enzyme-linked immunosorbent assay. Given the patient's history, physical examination results, and histologic findings, an atypical presentation of Darier disease was favored. Systemic corticosteroids, doxycycline, and niacinamide were discontinued, and the patient was started on acitretin 10 mg daily with topical triamcinolone 0.1% ointment. The patient was counseled regarding the genetic component of this condition, and a genetics referral was made. Unfortunately, the patient was lost to follow-up at this institution.

Darier disease is an uncommon, autosomal dominant condition (Schmieder and Rosario-Collazo, 2020). The vesiculobullous variant of Darier disease is particularly rare (Wang et al., 2019). Although vesiculobullous Darier disease shares histopathologic commonalities with PV, distinguishing between these etiologies is critical to provide patients with proper treatment. Our patient had carried an incorrect diagnosis of PV for several decades and was consequently treated at various institutions with courses of both topical and systemic steroids. Given her strong family history and examination findings, we questioned her initial diagnosis and decided to conduct repeat work-up. Her clinical morphology, negative immunofluorescence test results, and acantholysis with dyskeratosis on histology were diagnostic of vesiculobullous Darier disease rather than PV. Given her diagnosis, we discontinued her immunosuppressive regimen and started the patient on an oral retinoid, which has demonstrated efficacy in treating vesiculobullous Darier disease (Korman and Milani-Nejad, 2020).

Our case demonstrates the importance of performing a thorough patient history and examination and avoiding diagnostic anchoring. It also highlights a potential clinical and histopathologic pitfall in distinguishing between a rare variant of an inherited dermatologic syndrome and an autoimmune blistering disease.

Conflicts of interest

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Study approval

The author(s) confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies.

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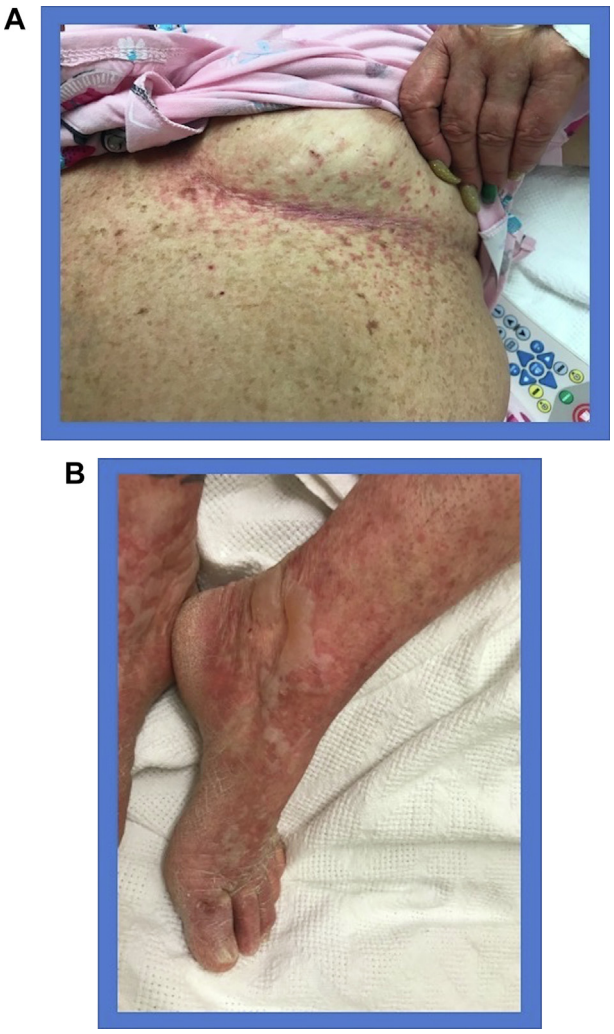

Fig. 1. (A) Hyperkeratotic erythematous papules and plaques in the inframammary region. (B) Flaccid bullae with erythema distributed on the patient's dorsal feet.

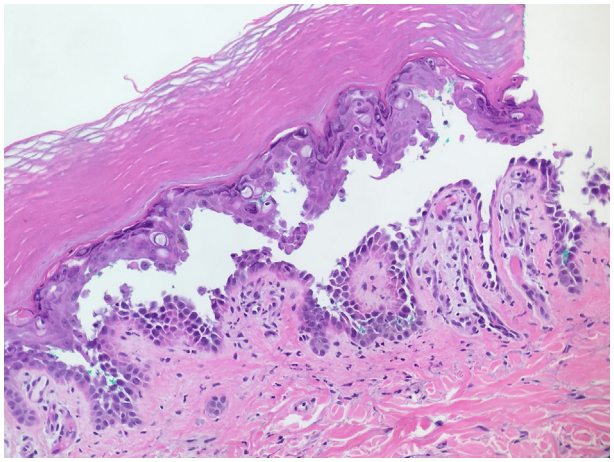

Fig. 2. Lesional skin on the right distal shin (hematoxylin and eosin stain, ×20).

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