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Authors
Sako, Eric Y
Worswick, Scott

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Extensive keloid formation after pemphigus vulgaris

Eric Y Sako MD, Scott Worswick MD

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Division of Dermatology, David Geffen School of Medicine at University of California, Los Angeles

Correspondence:
Scott Worswick, M.D.
Division of Dermatology, David Geffen School of Medicine at UCLA
200 Medical Plaza, Suite 450
Los Angeles, CA 90095
Email: SWorswick@mednet.ucla.edu
Phone: (310) 825-6911
Fax: (310) 794-7005

Abstract

Pemphigus vulgaris is an immunobullous disease characterized by intraepidermal blister formation. These blisters eventually rupture, leaving erosions that are slow to heal, often leaving hyperpigmented patches, but no scars. We describe a case of a 67-year-old man with pemphigus vulgaris who suffered severe keloidal scarring after the pemphigus lesions became infected. His keloids were treated with intralesional corticosteroids with some improvement. Pemphigus vulgaris, a process confined to the epidermis, may lead to scarring in predisposed individuals, particularly if infection occurs.

Keywords: pemphigus vulgaris, keloids, infection

Abbreviations: Pemphigus vulgaris (PV)

Introduction

Keloid scars are characterized by abnormal and exuberant collagen deposition following trauma to the dermis. These scars tend to overgrow the boundaries of the original injury, expand through invasion of surrounding dermis, and may form many months after the initial insult [1]. Although the exact mechanism by which keloids form is not known, this pathologic healing response is thought to begin within the dermis. Increased levels of dermal growth factors, dysregulation of extracellular matrix components, hyperactive fibroblast activity, and genetic background are all hypothesized to play a role [1,2]. Clinically, keloids can be pruritic, painful, and disfiguring, and sometimes are a challenge to treat [1].

Case synopsis

A 67-year-old man with pemphigus vulgaris (PV) diagnosed by biopsy and confirmatory indirect immunofluorescence presented to clinic with the complaint of painful new scarring as well as recurrent blisters on his body. The patient had been hospitalized 5 months earlier for a severe PV flare involving 15% body surface area. The course was complicated by secondary infection with meticillin-resistant staphylococcus aureus, treated with oral doxycycline and topical 2% mupirocin ointment. He was stabilized and discharged on a 60 mg prednisone taper, mycophenolate mofetil, topical clobetasol cream, and monthly intravenous immunoglobulin. Four months after his hospitalization, the patient noticed large scars forming within healed hyperpigmented...
patches that had previously been affected by PV. These scars were slowly growing, itchy, and painful. One week prior to presentation, the patient had been tapered to 10 mg of prednisone daily and had started to develop new blisters.

On exam, there were multiple confluent, polycyclic, purple to dark red plaques on his chest, arms, back, and buttocks (Figure 1). Numerous 1-3 cm, crusted, moist erosions were scattered on his face, lips, chest, back, and buttocks, with some developing on these aforementioned plaques (Figure 2). Multiple large, erythematous erosions with secondary alopecia were also found on his occipital scalp.

His exam was consistent with recurrent PV of the scalp and body arising amidst extensive PV-associated keloidal scars. The alternative diagnoses considered included deep fungal infection, dermatofibrosarcoma protuberans, and atypical mycobacterial infection. Biopsy was not performed given the consistent scar formation in places of past PV erosions and the distinct morphologic features. These keloids were treated with intralesional triamcinalone with resultant thinning. For the refractory PV, the patient was restarted on 60mg of prednisone and rituximab.

**Discussion**

Keloid formation following pemphigus is unusual. Because the process driving PV is largely confined to the epidermis, the lesions generally heal without scarring [3]. Rare case reports have described keloids forming after PV [4]. We surmise that the secondary infection of his PV erosions led to dermal injury, triggering a keloidal response in this predisposed patient. Cutaneous infection of PV lesions occurs in 10-16% of patients with the most common pathogen being staphylococcus aureus [5]. Risk factors for infection include widespread disease, diabetes, and systemic corticosteroids [5]. This case demonstrates that, despite appropriate treatment, PV may lead to significant scarring in patients prone to keloids. A history of keloids should prompt clinicians to be vigilant to recognize and quickly treat any secondary infection.

**References**