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CASE REPORT

Extensive keloid scarring in a patient with pemphigus vulgaris

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Abstract

Since pemphigus blisters are intraepidermal, scarring should induce at most a post-inflammatory hyperpigmentation. We describe a very atypical and unusual course of pemphigus vulgaris with extensive keloid formation despite high systemic steroids. This could be promoted by the severe flare of the disease, the delay of scarring, and the superinfection.

KEYWORDS

clinical dermatology, keloid, pemphigus vulgaris

1 | INTRODUCTION

Pemphigus vulgaris (PV) is the most common variant of pemphigus. It consists of intraepidermal blistering leading to flaccid bullae and mucosal and cutaneous erosions.[1] Keloid scarring in PV is uncommon. Since blisters are intraepidermal, scarring should induce at most a post-inflammatory hyperpigmentation. Only four cases of keloid scars following pemphigus have been reported.[2–5] Herein, we describe an unusual extensive keloid scarring following a severe flare of PV.

2 | OBSERVATION

A 47-year-old Caucasian woman with a 4-month history of PV presented with extensive keloid scarring at areas of prior PV erosions. Four months ago, she had been hospitalized for a severe PV flare with erosions affecting her trunk, and upper and lower limbs (Figure 1A). Oral, nasal, and conjunctival mucosa were also involved. In addition, nail damage was noted with onycholysis, onychomadesis, and necrotic paronychia (Figure 1B). The Pemphigus Disease Area Index was 55. The diagnosis of PV was confirmed by histological and immunological investigation showing intraepidermal blistering, supra-basal epidermal acantholysis, and inter-keratinocyte IgG deposits in direct immunofluorescence. ELISA was positive for anti-desmoglein 3 (120 U/ml) and anti-desmoglein 1 (180 U/ml). The erosions did not heal after 16 days of treatment with 1.5 mg/kg/day of prednisone-equivalent and became extensive and hemorrhagic. The patient was apyretic. The complete blood count revealed hyperleukocytosis and hyperneutrophilia. CRP level was 386 mg/L. Two days later, a greenish malodorous discharge covered almost all the erosions (Figure 2A). Cultures grew *Pseudomonas aeroginosa*. Thus, the diagnosis of ecthyma gangrenosum complicating a PV was retained. A dramatic improvement of the patient's skin erosions was observed 2 weeks after introducing an antibiogram-adjusted antibiotic therapy based on tazobactam and amikacin. After 2 weeks, the erosions had re-epithelialized with post-inflammatory hyperpigmentation, and the patient was discharged. We started oral steroids tapering without relapse. At the 4-month follow-up, extensive keloids were noted on previous superinfected PV sites (Figure 2B), for which intralesional corticosteroids were scheduled.

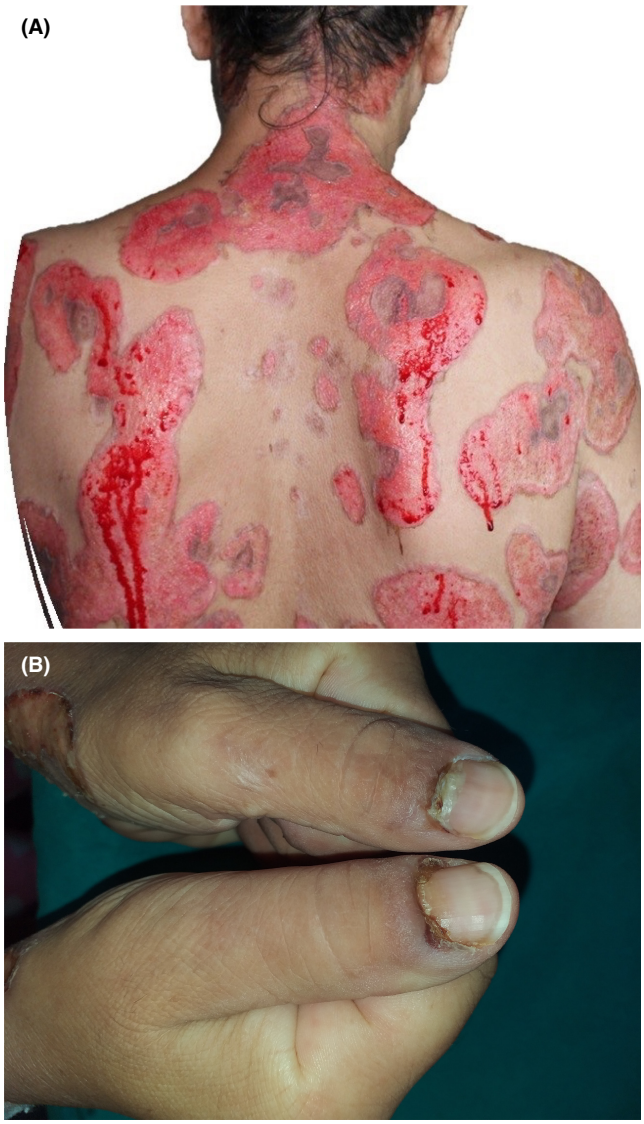

FIGURE 1 (A) Extensive hemorrhagic erosions on the trunk. (B) Onycholysis, onychomadesis, and necrotic paronychia

3 | DISCUSSION

Three main factors of keloids are known: infection, deep injury, and mechanical trauma.[6] So far, only four cases of keloid scars following pemphigus have been reported.[2–5] Gupta et al.[3] reported a case of uninfected PV erosions that healed with keloids despite high dose of systemic steroids. Shivaswamy et al.[4] described keloids in a patient whose pemphigus was treated, inter alia, with cyclosporin. The onset of keloids in this patient was attributed to cyclosporin. In the latest two cases, such as in our patient, extensive keloids appeared in patients whose pemphigus had been superinfected. The authors suggested that secondary infection induced keloids via dermal damage. However, the paucity of cases of keloids following PV flares contrasts with the frequency of superinfections which are common in pemphigus, suggesting the presence of other unknown factors.[2,5]

Our case is notable due to the severity of the PV flare, which was refractory to conventional treatment and then complicated by ecthyma gangrenosum. Re-epithelialization took more than 1 month and ended with extensive keloids after 3 months. It is reasonable that superinfection led to deep intradermal inflammation and keloid formation. In fact, keloids occurred in previously superinfected PV areas. In this context, there are reports of keloid onset following herpetic infections, with the keloids involving previously infected areas.[7] An alternative hypothesis is that erosions in our patient took exceptionally long to heal, which maintained a long inflammation and scarring process with a chronic fibroblast activation. This does not exclude the potential role of apoptotic cells and increased levels of tumor necrosis factor-alpha, found in PV, that may contribute to the dysregulation of fibroblasts, and lead to keloids in susceptible patients.[8] Intralesional steroids are generally proposed in patients

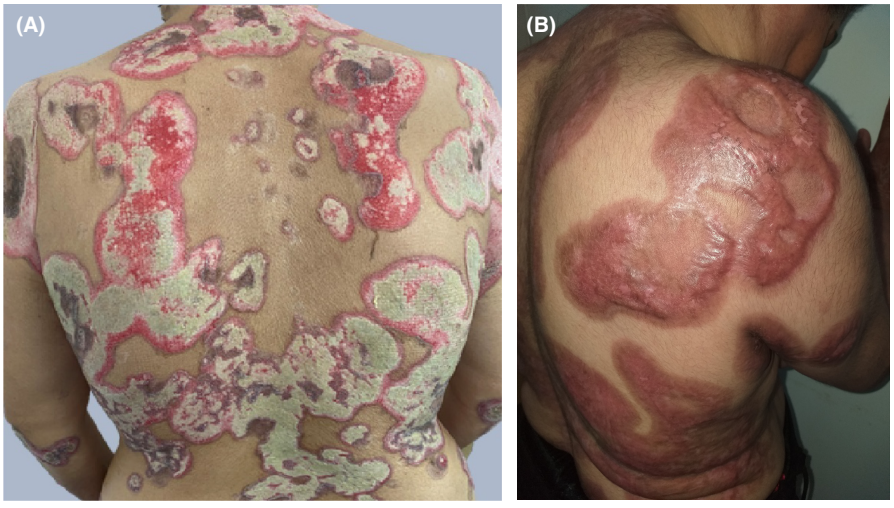

FIGURE 2 (A) Greenish discharge covering the erosions. (B) Extensive keloids on previous superinfected areas

with keloids, yet our patient is under high systemic doses of steroids, making this alternative debatable as we can opt for compression clothing and active monitoring.

4 | CONCLUSION

This case is being reported to highlight a very atypical and unusual course of PV with extensive keloid formation despite high systemic steroids. This could be promoted by the severe form of PV, the delay of scarring, and the superinfection. Moreover, treatment in such cases may be challenging since patients are usually under systemic steroids making intralesional steroids contestable.

AUTHOR CONTRIBUTIONS

Dorsaf Elinkichari is the guarantor of the content of the manuscript, including the data and analysis. Anissa Zaouak contributed to the acquisition of data, conception, andinterpretation of information and gave final approval of the version to be submitted. Amal Chamli revised data critically for important intellectual content. Houda Hammami and Samy Fenniche contributed to the interpretation of data and revisionof the manuscript.

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None.

CONFLICT OF INTEREST

None.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

The patient has given written informed consent to the publication of his case details.

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