Koebner Phenomenon in Epidermolysis Bullosa Acquisita: Letter to the Editor

Epidermolizis Bülloza Akizada Köbner Fenomeni

İbrahim KÖKÇAM, MD,[a]
Nusret AKPOLAT, MD[b]

Departments of [a]Dermatology, [b]Medical Pathology, Fırat University Medical Faculty, Elazığ

Geliş Tarihi/*Received:* 15.05.2008
Kabul Tarihi/*Accepted:* 20.05.2008

Yazışma Adresi/*Correspondence:*
İbrahim KÖKÇAM, MD
Fırat University Medical Faculty, Department of Dermatology, Elazığ
TÜRKİYE/TURKEY
ibrahimkokcam@hotmail.com

Dear Editor,

Epidermolysis bullosa acquisita (EBA) is a rare chronic autoimmune blistering disease that affects the skin and mucous membranes.[1,2] Koebner's phenomenon is encountered when the typical features of a dermatosis are observed on a skin part previously subject to friction or trauma. This phenomenon was reported in a variety of dermatoses.[3,4]

To our knowledge, this is the first reported case of EBA manifesting with a Koebner-like phenomenon due to scratching.

A 22-year-old man presented to the Dermatology Clinic of Fırat University Hospital with recurrent, intensely itchy and crusting vesiculobullous lesions with a duration of 10 years. Personal and family history revealed no specific data. Examination revealed multiple ulcerated and crusted lesions, erosions, tense bullous and vesicular lesions, and milia formations distributed mainly over the flexural aspects of the lower extremities. Moreover, he had linear vesicular lesions resembling Koebner phenomenon (Figure 1). There were no mucosal involvements. Very few lesions were present on the trunk, hands and feet. In addition, there were dystrophic changes of the toe and finger nails (Figure 2). Systemic examination revealed no abnormal findings. Routine hematological and biochemical parameters were within normal limits. Histopathological examination of the biopsy materials revealed a subepidermal blister formation. Edema and perivascular mild chronic inflammatory infiltrations, with lymphocyte predomination were noted on the superficial dermis. There was no eosinophilic infiltration. Direct immunofluorescence (DIF) study of perilesional skin showed linear IgG and C3 deposits at the basal membrane level. The clinical and histopathologic findings were consistent with the diagnosis of EBA.

EBA is a rare, chronic, autoimmune blistering disease that usually affects middle-aged to elderly adults.[1,2,5] Three clinical variants of EBA exist. In the non-inflammatory mechanobullous form, blisters arise on a non-erythematous base and heal with scar and milia formation. Nail dystrophy may

FIGURE 1: Blisters, erosions, scarring and linear vesicular lesions on the lower extremities.

FIGURE 2: Blisters, erosions, scarring and linear vesicular lesions on the lower extremities.

be present in the majority of the cases. The inflammatory form is characterized by blisters and vesicles on erythematous skin. This inflamatory vesiculobullous eruption shows marked similarities with bullous pemfigoid. The blisters and vesicles may be very pruritic. In this form, scarring and milia formations are less prominent compared to the previous one. Typical sites are the trunk and flexural aspects of the extremities. The blisters frequently remain intact and are not enlarged with subtle pressure. A third type clinically resembles cicatricial pemfigoid and is characterized by prominent mucosal involvement.[2,6]

The histology of an EBA lesion demonstrates subepidermal blister formation. The infiltrate within the dermis can be variable, and in some patients, it is not present. DIF examination of the perilesional tissue reveals a linear deposition of IgG and C3 at the dermoepidermal junction in most cases. DIF using salt split skin demonstrates binding of antibodies from the sera of EBA patients to the dermal side of the split. The main histological differential diagnosis is bullous pemphigoid, which is characterized by moderately dense eosinophilic and other inflammatory cells.[1,2,6,7] Our patient had subepidermal blister with perivascular lymphocytic infiltration without eosinophils. These findings supported the diagnosis of EBA. The dilemma in the diagnosis of our case was the lack of salt-split skin study for bullous pemfigoid considered in the differential diagnosis because of technical inadequacy. Our clinical clues were the presence of milia and nail dystrophy in our patient.

Koebners' phenomenon was reported in a variety of dermatoses.[2,3] It is rare in chronic bullous diseases.[2] However, in the available literature, our presentation is the first report of Koebner phenomenon occurring in EBA patients. The exact pathogenesis of the Koebner phenomenon remains unknown. Although mechanical trauma like pruritus as a precipitating factor of blister formation in bullous pemphigoid was previously reported, the exact pathogenesis is still unclear.[8] Some authors consider it as the result of different forms of epidermal damage in predisposed patients, with the induction of antigen exposure in the context of subclinical pemphigoid followed by activation of the corresponding autoimmune process.[9-11] It is likely that in our case the scratching secondary to the intense pruritus may act as a trigger factor for the development of the linear vesicular lesions.

In conclusion, we report that a Koebner phenomenon-like eruption may occur in EBA. Therefore, the differential diagnosis in patients with Koebner phenomenon-like vesiculobullous eruptions should also include EBA.

REFERENCES

1. Sitaru C, Zillikens D. Mechanisms of blister induction by autoantibodies. Exp Dermatol 2005;14:861-75.
2. Hallel-Halevy D, Nadelman C, Chen M, Woodley DT. Epidermolysis bullosa acquisita: update and review. Clin Dermatol 2001;19:712-8.
3. Thappa DM The isomorphic phenomenon of Koebner. Indian J Dermatol Venereol Leprol 2004;70:187-9.
4. Boyd AS, Neldner KH. The isomorphic response of Koebner. Int J Dermatol 1990;29:401-10.
5. Yeh SW, Ahmed B, Sami N, Razzaque Ahmed A. Blistering disorders: diagnosis and treatment. Dermatol Ther 2003;16: 214-23.
6. Smoller BR, Kohler S. Subepidermal vesicular dermatitis. In: Barnhill RL, Crowson AN, eds. Texbook of Dermatopathology. 2nd ed. New York: McGraw-Hill Medical Pupb; 2004.p. 167-96.
7. Gammon WR, Briggaman RA, Woodley DT, Heald PW, Wheeler CE Jr. Epidermolysis bullosa acquisita--a pemphigoid-like disease. J Am Acad Dermatol 1984;11(5 Pt 1):820-32.
8. Dahl MG, Cook LJ. Lesions induced by trauma in pemphigoid. Br J Dermatol 1979;101:469-73.
9. Pardo J, Rodrguez-Serna M, Mercader P, Fortea JM. Localized bullous pemphigoid overlying a fistula for hemodialysis. J Am Acad Dermatol 2004;51:131-2.
10. Parslew R, Verbov JL. Bullous pemphigoid at sites of trauma. Br J Dermatol 1997;137:825-6.
11. Macfarlane AW, Verbov JL. Trauma-induced bullous pemphigoid. Clin Exp Dermatol 1989;14:245-9.