Pemphigoid Excoriee: An Unusual Type of Bullous Pemphigoid Presenting with Köbner Phenomenon¶

PEMFİGOİD EKSKORYE: KÖBNER FENOMENİ İLE BAŞVURAN ALIŞILMAMIŞ BİR BÜLLÖZ PEMFİGOİD TİPİ

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Summary

Bullous pemphigoid is an acquired autoimmune subepidermal blistering disease that most commonly affects elderly persons. Classically, pruritic blisters develop on normal or erythematous skin. Although its onset is generally spontaneous, ultraviolet light, topical or systemic medications can induce bullous pemphigoid as well. The induction of pemphigoid lesions by trauma seems largely to have little attention in the literature. We describe a patient who developed an unusual pattern of bullous pemphigoid which was localized only to the areas of chronic excoriation. There is only one report of this atypical case in literature and it was called as "Pemphigoid excoriée" by the authors.

Key Words: Bullous pemphigoid, Pemphigoid excoriée, Trauma-induced bullous pemphigoid, Köbner phenomenon

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Özet

Büllöz pemfigoid genellikle yaşlı kişilerde görülen akkiz, otoimmün, subepidermal büllerle karakterize bir hastalıktır. Klasik olarak, normal veya eritemli zeminde kaşıntılı büller gelişir. Başlangıç genellikle spontan olmasına rağmen, ultraviyole ışığı, topikal ya da sistemik ilaçlar büllöz pemfigoidi indükleyebilir. Literatürde, travma ile büllöz lezyonların oluşumu az sayıda çalışmada dikkat çekmektedir. Burada sadece kronik ekskoryasyon alanlarına lokalize büllöz lezyonların oluştuğu bir büllöz pemfigoid olgusu sunulmaktadır. Sunulan olgumuza benzer literatürde tek yayın bulunmaktadır ve yazarlar bu atipik kliniği "Pemfigoid ekskorye" olarak tanımlamışlardır.

Anahtar Kelimeler: Büllöz pemfigoid, Pemfigoid ekskorye, Travmaya bağlı büllöz pemfigoid, Köbner fenomeni

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Bullous pemphigoid (BP) is a chronic immunobullous disease of the elderly. Classically, tense and pruritic blisters develop on the skin. Recently, the advent of direct and indirect immunofluorescense testing has expanded the study of BP. There are many reported atypical special forms of BP some of which are well defined in addition to the idiopathic form in literature (1,2). We describe herein a 73 year-old woman who developed an unusual pattern of BP which was localized to the areas of chronic excoriation. There is only one report of this atypical case in literature and it was called as "Pemphigoid excoriée" by the authors (3).

Case report

A 73-year-old woman referred to our clinic with widespread bullous lesions and severe pruritus on her body for six months. Her family history was noncontributory. She had hypertension for one year. Physical examination revealed widespead ulcerated and crusted lesions ranging from 5 to 20 mm in diameter on her arms, trunk and legs, coexisting with bullous lesions that were small and tense. She had generalized pruritus and the lesions were remarkably confined to the scratched areas. She didn't have any lesions over her interscapular area and her back (Fig 1). She had linear excoriated lesions resembling Köbner phenomenon (Fig 2). There were no mucosal involvements. A biopsy specimen of excoriated lesion showed a subepidermal blister containing neutrophils and eosinophils with perivascular eosinophils and lymphocytes (Fig 3). Direct immunofluoresence microscopy (DIF) revealed deposition of linear IgG and C3 along the basement membrane (Fig 4). Immunofluoresence examination of salt-split patient's skin showed IgG binding to the roof of the blister. Bullous pemphigoid was diagnosed and treatment with prednisolone 1.5 mg/kg daily was initiated. Since the lesions persisted after two weeks of therapy, methotrexate 10 mg weekly was added. The lesions improved and pruritus decreased in the next two weeks and prednisolone dose was tapered to 20 mg daily. Methotrexate was stopped after four weeks.

Discussion

Bullous pemphigoid (BP) is a chronic disease which is characterized by subepidermal blisters. The disease may

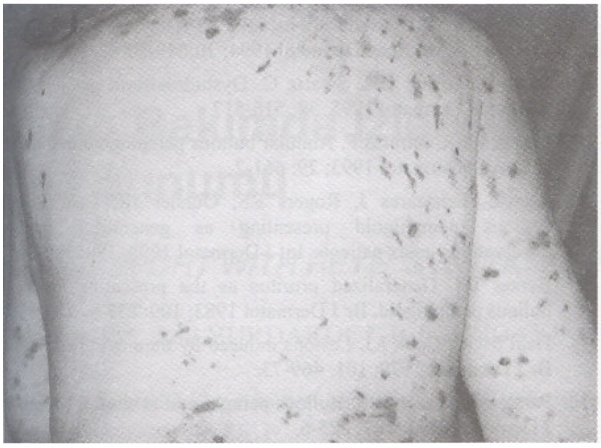

Figure 1. Excoriations are seen to spare the upper back.

Figure 2. Clusters of small tense blisters and linear excoriated lesions on the thigh.

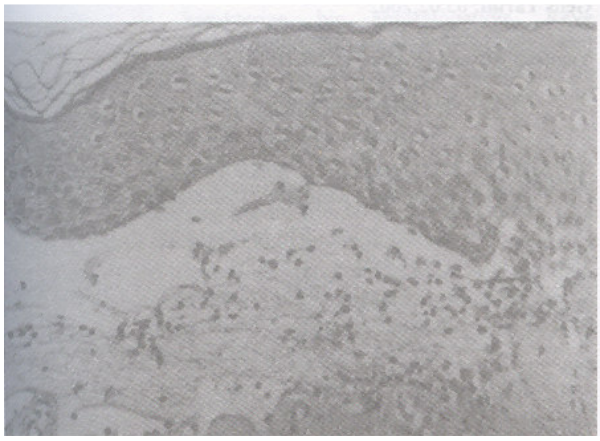

Figure 3. Skin biopsy specimen of excoriated lesion: subepidermal bulla containing neutrophils and eosinophils with perivascular eosinophils and lymphocytes (H&E, 40).

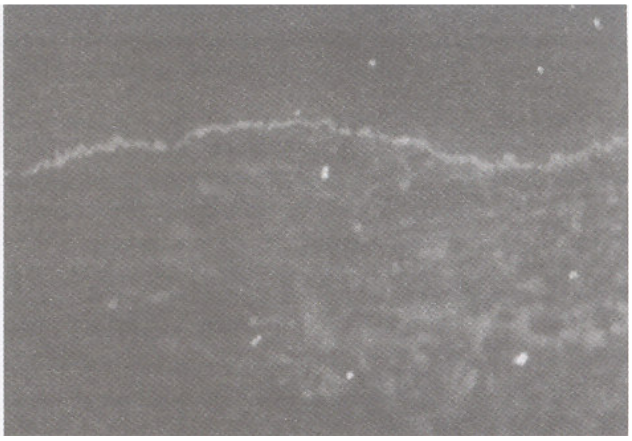

Figure 4. Direct immunofluorescense: Linear IgG deposition at the dermoepidermal junction.

typically involve the whole body. Tense vesicles and bullae arise spontaneously either on normal or inflammed skin.There may be some atypical variants of BP such as erythematous and edematous, vesicular, localized, seborrheic, vegetating, urticarial, atypical, subclinical, erythrodermic, dishidrosiform, nodular, annular and polimorphic. These variants can be diagnosed reliably only after histological and immunopathological examination (1,4,5-7).

Generalized itching in the elderly can be result of a variety of systemic and cutaneous disorders, but many elderly patients remain undiagnosed. However, severe pruritus is a characteristic symptom of BP and may precede the development of bullous lesions (1,8). The clinical presentation of BP with severe and persistent pruritus was described as “pruritic pemphigoid” (7).

Mechanical trauma like pruritus as a precipitating factor of blister formation in BP has been previously reported. The cutaneous lesions of BP induced by mechanical trauma are usually strictly located at the site of the damaged area with little outside spreading (9). In such cases, diagnosis may be confused with epidermolysis bullosa acquisita. It has been suggested that chronically self-inflicted trauma with pruritus injure the epidermis and dermis and, as a result the injured dermo-epidermal junction has exposed relatively sequestered constituents of the epidermal basement membrane and has encouraged an autoimmune response (3,10). The damaged epithelium may act as a stimulus for anti-BMZ antibody production (10,11). Except mechanical trauma, radiotherapy and ultraviolet light have been reported as a trigger factor for the development of BP (10).

We suggest that this case is a different clinical variant of BP. The scratching was a dominant symptom and blisters were dramatically confined to scratched areas in our patient. Allan et al (3) had described a case with BP which had atypical presentation like our case. They called this variant as "pemphigoid excoriée".

Clearly in a majority of patients with BP, trauma-induced lesions are not of clinical importance. There is a close analogy with psoriasis in which trauma-induced lesions (Köbner phenomenon) may often be found on careful examination but rarely dominate the clinical picture (12). As a result, mechanical trauma like pruritus is a precipitating factor of blister formation in BP. This unusual trauma-induced variant of BP may initially be easily overlooked. Elderly patients with severe and persistant unexplained pruritus should go through immunofluorescense testing to exclude BP as the cause of the generalized pruritus.

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