



Bullous Pemphigoid Mimicking Discoid Lupus Erythematosus: A Case Report

LETTER TO THE
EDITOR

Derya Uçmak¹, Zeynep Meltem Akkurt¹, Bilal Sula¹, Gurbet Acar¹, Ulaş Alabalık², Mustafa Arıca¹

Bullous pemphigoid, which rarely includes mucous membranes, is a chronic, autoimmune, subepidermal, and bullous skin disease. Bullous pemphigoid is characterized with the presence of specific immunoglobulin G (IgG) autoantibodies against BP230 (BP AG1) and BP180 (BP AG2), which are hemidesmosomal bullous pemphigoid antigens (1). Taut vesicles and bullae may occur spontaneously on normal and inflamed skin. Before the development of bullae-blisters, prodromal phase itching, lasting a few weeks or even a few months, may occur alone or may accompany excorie, eczematous, papular, or urticaria-like lesions. While non-bullous skin symptoms continue, depending on the clinical symptoms of the lesion, some clinical variants may emerge in some patients. These are prurigo-like, erythroderma-like, ecthyma gangrenosum-like, intertrigo-like, and toxic epidermolysis-like bullous pemphigoid (2). All of these variants also have the same histological and immunological characteristics of the classical form of bullous pemphigoid (1).

In this article, a male case who was diagnosed with bullous lesions that had started with discoid lupus erythematosus (DLE)-like lesions in the face that then spread to the trunk and back will be presented.

CASE REPORT

One year ago, a 63-year-old male patient was admitted to our clinic with color change symptoms in the right frontotemporal region, pinkish in the edges and purplish in the center. There was no pain, but itching was accompanying his complaint. The dermatological examination revealed a 6-cm-diameter plaque in the frontotemporal region that was livedoid in color with a darker color in the center (Figure 1). There was a 3-cm linear erythematous lesion in the right lateral cheek. There was no involvement in the oral and ocular mucosa. Apart from these lesions, his dermatological examination was normal. The case was biopsied with DLE, lichenoid drug eruption, and chronic actinic dermatitis pre-diagnosis, and his biopsy showed that there were orthokeratosis on the surface, focal parakeratosis, thinning on the epidermis, mild spongiosis in the basal layer, exocytosis of lymphocytes in places, and focal vacuolar degeneration. The patient was clinically and histopathologically diagnosed with discoid lupus erythematosus. System examination of the case revealed that there was no involvement, and antinuclear antibodies and laboratory examinations were within normal limits. The patient was given topical steroid treatment and was told to visit the clinic for follow-up care. The case had no symptoms for a year but was readmitted to our clinic afterwards with plaque lesions that had minimal vesicles on the edges of a slightly erythematous base on his back, belly, and sides of the body (Figure 2). Simultaneously, there was a prominence of lesions in the temporomandibular regions and newly formed plaque lesions on the other cheek that were similar to the older ones. In the biopsy performed on his back, we observed dissociation in the subepithelial region of the tissue showing acanthosis and spongiosis on the surface and eosinophil formation and, in places, neutrophil formation on the epithelium that was mostly ulcerated in the dissociation region (H&E, 100x) (Figure 3). The patient was diagnosed with bullous pemphigoid and given a 60 mg/kg-dose of methylprednisolone treatment. Two weeks later, the lesions of the patient in remission regressed partially; 3 months later, the lesions regressed completely, leaving postinflammatory hyperpigmentation (Figure 4).

Bullous pemphigoid is a chronic, acquired, autoimmune, subepidermal bullous disease that is mostly observed in the elderly and characterized by autoantibodies and complement storage in the basement membrane zone. The most characteristic feature of bullous pemphigoid is that the bullae are usually accompanied by severe itching and develop on erythematous or normal-looking skin (1, 3). In our case, the back and body lesions were shape slightly like erythematous plaques and were accompanied by pronounced itching.

¹Department of Dermatology,
Dicle University Faculty of
Medicine, Diyarbakır, Turkey

²Department of Pathology,
Dicle University Faculty of
Medicine, Diyarbakır, Turkey

Submitted
06.01.2013

Accepted
14.11.2013

Correspondance
Derya Uçmak MD,
Department of Dermatology,
Dicle University Faculty of
Medicine, Diyarbakır, Turkey
Phone: +90 412 248 80 01
e.mail:
ucmakderya@gmail.com

Presented in the "7th Aegean
Dermatology Days" in the book
of abstracts

©Copyright 2014
by Erciyes University School of
Medicine - Available online at
www.erciyesmedj.com



Figure 1. A 6-cm-diameter plaque in the frontotemporal region that is livedoid in color with a darker color in the center

In about 30% of bullous pemphigoid disease cases, the disease starts in one part of the body and then spreads. The bullae may develop in any part of the body but are frequently observed at the bottom of the abdomen and in the inguinal region and flexor surfaces of the extremities. Unlike this dominant settlement, bullous pemphigoid that starts on the face is very rare (1).

Based on the positive immunofluorescence findings, many clinical subtypes of the disease have been identified. The clinical classification of bullous pemphigoid is divided into two: generalized and localized. Whereas generalized types are classified as bullous, urticaria, vesicular, vegetans, and nodular, the peritibial and dyshidrosiform ones are classified as non-cicatricial localized, and the Brunsting-Perry type is classified as cicatricial localized (1). Apart from these clinical variants, erosive bullous pemphigoid and drug-induced bullous pemphigoid were also added (3).

Despite the differences in the appearance of the lesions in the clinical types of bullous pemphigoid that are seen less frequently, most patients may be diagnosed with the help of positive findings obtained with typical clinical findings, histological features, and, most importantly, DIF analysis (1).

It is best to section from newly formed bullae on the inflamed skin and perform staining with hematoxylin and eosin on it. Histologically, with eosinophils, there are subepidermal bullae in the dermis and bulla cavity. However, bulla in pemphigoid may be formed on inflamed skin or on skin that shows inflammation. In the histopathology of the lesion, there is intense inflammatory infiltrate in the



Figure 2. Plaque lesions having minimal vesicles on the edges of the slightly erythematous base on his back

subepidermal region consisting of lymphocytes, neutrophils, and especially eosinophils. There is perivascular infiltrate consisting of lymphocytes and eosinophils (1). Although it resembles other subepidermal bullous diseases clinically and histologically, it is important in terms of distinguishing direct immunofluorescence disease. In immunofluorescence studies, in the epidermal zone of the basement membrane, to observe the storage of IgG (80%) and C3 (100%) in a band shape is utilized (1, 4). Our case was diagnosed with bullous pemphigoid using current clinical and histopathological findings.

Although the morphology and immunopathology of autoimmune bullous diseases are very similar, the progression of the disease, age of onset, the presence and severity of mucosal involvement, and whether there is a scar or not play an important role in the diagnosis and differential diagnosis (4). Direct immunofluorescence and salt-split indirect immunofluorescence methods are important to distinguish bullous pemphigoid from other bullous diseases. Furthermore, cellulitis, bacterial and viral infections, insect bites, and erythema multiforme should be considered in the differential diagnosis (4). The first lesion of our patient was clinically a discoid plaque type purplish livedoid in color with a slight squam on it. The case was first diagnosed with DLE, and his lesions regressed through topical steroid use, but after a 1-year period without having any lesions, the case suffered from lesion recurrence not only in the same area but also in other parts of the body.

The efficacy of systemic steroids in bullous pemphigoid treatment has been proven. As soon as the patient started using the

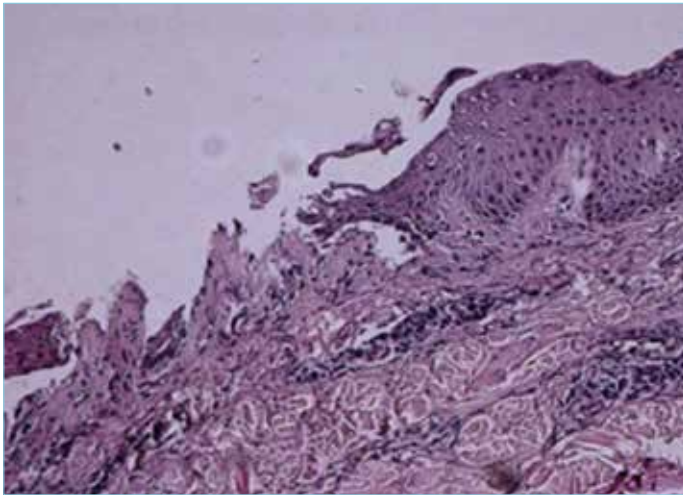


Figure 3. Dissociation in the subepithelial region and dissociation region of the tissue showing slight acanthosis and spongiosis on the surface (H&E, 100x)



Figure 4. The appearance of postinflammatory hyperpigmentation plaque and lesion 3 months later

drug, within 1-4 weeks, it was observed that bullae and inflammation showed rapid suppression; however, the dose should then be gradually reduced (4). For severe involvement, 0.75-1 mg/kg is suggested, in moderate diseases, 0.5 mg/kg is suggested, and

for mild or localized diseases, 0.3 mg/kg is suggested (4). Due to complications of steroids, the steroid dose and duration should be minimized, and therefore, other immunosuppressive drugs might be added to the treatment. Treatment should aim to take the symptoms under control with minimum side effects (1). We started a 60 mg/day-dose of methylprednisolone treatment in our case, who had plaque lesions livedoid in color and bullous lesions on his back and front part of the body, and we achieved partial recovery after the end of the second week. The dose was reduced gradually, and we stopped the treatment. Lesions regressed completely after a 3-month treatment, leaving postinflammatory hyperpigmentation.

Recently, an 83-year-old case with cicatricial pemphigoid that mimicked DLE was reported in a study conducted by Das et al. (5), and this is the first case report showing that cicatricial pemphigoid may mimic DLE clinically. At first, due to the clinical findings, our case was also diagnosed with DLE, but it should be remembered that bullous pemphigoid might be experienced with clinically DLE-like lesions, as well. Clinically, a bullous pemphigoid type resembling DLE has not been defined in the literature yet. Our case was presented as a rare case in terms of being the first of its kind.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Authors' Contributions: Conceived and designed the experiments or case: DU. Performed the experiments or case: ZMA. Analyzed the data: BU. Wrote the paper: DU, BU, GA. All authors have read and approved the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Ghohestani RF, Novotney J, Chaudhary M, Agah RS. Bullous pemphigoid: from the bedside to the research laboratory. *Clin Dermatol* 2001; 19(6): 690-6. [\[CrossRef\]](#)
2. Schmidt E, della Torre R, Borradori L. Clinical features and practical diagnosis of bullous pemphigoid. *Dermatol Clin* 2011; 29(3): 427-38. [\[CrossRef\]](#)
3. Walsh SR, Hogg D, Mydlarski PR. Bullous pemphigoid: from bench to bedside. *Drugs* 2005; 65(7): 905-26. [\[CrossRef\]](#)
4. Venning VA, Taghipour K, Mohd Mustapa MF, Hight AS, Kirtschig G. British Association of Dermatologists' guidelines for the management of bullous pemphigoid 2012. *Br J Dermatol* 2012; 167:1200-14. [\[CrossRef\]](#)
5. Das S, Blanco G, Ahmed A, Tran KT, Matthews LA, Pandya AG. Cicatricial pemphigoid of the scalp mimicking discoid lupus erythematosus. *J Am Acad Dermatol* 2011; 65(4): 886-7. [\[CrossRef\]](#)