

Case Report

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Sudden Onset Erythrodermia, Generalized Edema and Bullae Formation in A Male Patient With A 20 Years History of Psoriasis

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Abstract

Observation: Appearance of bullous pemphigoid in patients with preexisting psoriasis vulgaris is a rare clinical entity. We present a 70 years old male patient with sudden onset erythrodermia, generalized edema and tense bullae formation in the setting of a 20 years history of psoriasis. Rapid clinical response was observed after systemic corticosteroid therapy was initated along with methotrexate therapy.

Introduction

Bullous pemphigoid is a disease of elderly, which may be the presenting sign of an underlying occult visceral malignancy. The appearance of bullous pemphigoid in patients previously diagnosed with psoriasis is a rare clinical entity. Here we present an elderly male patient with coexisting psoriasis and bullous pemphigoid that were accompanied by generalized edema.

Case Report

A 70 years old male patient presented to the outpatient clinic with erythrodermia. He also complained of fever, chills, malaise, pruritus and scrotal swelling. He has been diagnosed with psoriasis vulgaris 20 years ago, was treated with topical steroids and emollients and never received systemic treatment. The disease was never documented by a biopsy. He reported that, within the past two weeks, red pruritic plaques appeared with a widespread distribution. His past medical history was unremarkable and had no regular medications. Upon the physical examination, he had infiltrated, desquamated, erythematous plaques and excoriation marks all over the body. Scrotal edema, pretibial edema and ascites were also present. The patient was admitted with the diagnosis of erythrodermia.

The initial biochemistry revealed hypoalbunemia, anemia, increased erythrocyte sedimentation rate and increased lactate dehydrogenase. He was consulted to the internal medicine and urology departments and albumin replacement was initiated. Further more, thoracic, abdominal and pelvic CTs with intravenous and oral contrast were taken with the suspicion of an underlying malignancy given the age and the sudden presentation of the patient. The CTs revealed multiple para-aortic lymphadenopathies that were concluded to be dermatopathic by the internal medicine consultant. Thus it was shown that there was no underlying malignancy. Generalized edema regressed after albumin replacement. 1st generation sedating H1 antihistamines were given orally three times a day, yet the pruritus was refractory.

J Turk Acad Dermatol 2019; 13(1): 19131c2.

On the third day of his admission, widespread flaccid bullae appeared on the abdomen and extremities, both on the erythematous plaques and normal skin (**Figures 1,2,3 and 4**). Biopsies were taken from the bullae and plaques with the suspicion of bullous pemphigoid. The pathology specimen taken from a plaque revealed psoriasis vulgaris; and the specimen taken from a bullae revealed bullous pemphigoid. Direct immunofluorescence testing supported bullous pemphigoid as well. For that reason, the patient was diagnosed with coexisting psoriasis and bullous pemphigoid.

Systemic corticosteroid treatment was initiated at 48mg/day. New bullae formation ceased at the 5th day of corticosteroid therapy. Methotrexate treatment was initiated at 20 mg/week as well. The patient's symptoms subsided and erythrodermia regressed. Corticosteroid dose was tapered after methotrexate was initiated.

Discussion

The coexistence of psoriasis vulgaris and bullous pemphigoid is observed rarely. It was first observed by Bloom in 1929 [1]. Until now,



Figure 1. Intact and eroded vesicles along with excoriated papules on the forearm

forty patient presenting with coexisting psoriasis and bullous pemphigoid has been reported. These patients have an average age of 63, ranging from 47 to 86. Men were affected 3 times more common than women. The average time interval between the onset of psoriasis and onset of bullous pemphigoid was 20 years [2]. Similar to the cases reported before, our patient was also male and has had psoriasis for 20 years; however, unlike the previously reported range, our patient was a little older, which prompted us with the possibility of a malignancy.

It was previously postulated that anti-psoriatic therapies may alter the basement membrane and result in immunologic responses that leads to the development of bullous pemphigoid: phototherapy and topical tar are of particular concern [2]. Our patient has only received topical corticosteroids and emollients, which precludes this possibility. Furthermore, it was recently shown that interleukin 17 (IL-17) plays a role not only in the pathogenesis



Figure 2. Intact bulla on the right foot with a background of edema and erythema

J Turk Acad Dermatol 2019; 13(1): 19131c2.

Figure 3. Intact and eroded vesicles on an erythematous background on the bilateral pretibial areas

of psoriasis vulgaris but also in the pathogenesis of bullous pemphigoid; thus targeting IL-17 can resolve both pathologies. IL-17 also has a crucial role in metabolic syndrome, which frequently coexists with psoriasis vulgaris. A case with concomitant psoriasis vulgaris, bullous pemphigoid and diabetes mellitus was reported in 2016 [**3**].Unlike this case, our patient had no pre-existing diabetic comorbidity.

The treatment possibilities targeting both pathologies are systemic corticosteroids, methotrexate, acitretin, dapsone, azathioprine and mycophenolate mofetil [2,4]. Systemic corticosteroids hold the risk of pustular transformation in psoriasis, therefore should be given and tapered cautiously. Etanerept, which is tumor necrosis factor alpha (TNF-alpha) blocker, can be used if the patient is refractory to conventional therapies [4]. Agents targeting IL-17 (i.e. secukunumab) holds future promise for refractory patients as well [3].

Conclusion

Coexistence of psoriasis vulgaris and bullous pemphigoid is a rare clinical entity. The un-

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Figure 4. Intact and eroded vesicles on posterior tibia

derlying pathogenesis of this situation is not yet understood clearly. Corticosteroids and other immune-modulating drugs are beneficial in the treatment of both diseases. Our case was distinct in that generalized edema also accompanied the abrupt clinical onset, which prompted us with the possibility of an underlying visceral malignancy. Yet, no malignancy was detected and the patient's symptoms resolved with systemic corticosteroids and methotrexate therapy.

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