Dermatology Case Reports

Case Report Open Access

Coexistence of Pemphigus Vulgaris and Pemphigus Foliaceus in the Same Patient at the Same Time

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Abstract

Pemphigus is an autoimmune blistering disease and includes two classical forms. These forms are Pemphigus vulgaris (PV) and Pemphigus foliaceus (PF). These two subtypes of the disease are related with different auto-antibodies and show different clinical features. Rare cases of simultaneous presence of PV and PF have been presented in the literature. Here, we report the case of a 62-year-old woman who developed PV and PF lesions at the same time.

Keywords: Pemphigus disease; Simultaneous presence; Two subtypes

Introduction

Pemphigus is an autoimmune blistering skin disease resulting from a loss of keratinocyte cell adhesion mediated by autoantibodies against desmoglein 1 (Dsg1) and/or desmoglein 3 (Dsg3). Pemphigus disease that can be divided into two major subtypes by the result of these autoantibodies reactivity: Pemphigus vulgaris (PV) (target antigen is Dsg3) and pemphigus foliaceus (PF) (target antigen is Dsg1). The PV antigen, which is defined by autoantibodies from PV patients, has been characterized as a 130 kDa glycoprotein (desmoglein 3), whereas the PF antigen is a 160 kDa or 150 kDa desmosomal glycoprotein (desmoglein 1) [1-12]. These two types of the disease present some different clinical features. For instance, PV is characterized with oral and cutaneous erosions but oral mucosal lesions are only rarely seen in PF. We present a patient who has these two subtypes of the disease at the same time.

Case Report

A 62-year old woman who lives in Konya from Turkey was admitted to our dermatology clinic with 1-month history of oral mucosal erosions 4 years ago. Skin examination revealed multiple blisters on the bilateral buccal mucosas and the soft palate. A punch biopsy specimen from the oral mucosa revealed suprabasilar acantholysis in the epidermis. Direct immunofluorescence showed intercellular deposits of IgG in the lower epidermis confirming the clinical diagnosis of PV. Past medical history of the patient revealed that she was affected by hypertension, diabetes mellitus type-2, hyperlipidaemia and ischemic stroke. Methylprednisolone I.V. at the initial dose (80 mg per day) and Azathioprine P.O. (150 mg per day) was given to the patient. Cause of development steroid-induced myopathy and confusion, confabulation, amnesia like neurological symptoms (she has complained weakness mainly to the proximal muscles of the upper and lower limbs) then methylprednisolone dose was gradually reduced to 16 mg per day in 3 months. In a few months, patient's lesions were spread into the scalp and her chest. She was suffering from pruritus and crusts on healing lesions. Azathioprine was stopped and Cyclosporine P.O. 200 mg per day was given for the adjuvant therapy. Topical corticosteroids and emollients were also added. She was seemed in the visits periodically. Her BUN and creatinine levels was increased and blood pressure values were uncontrollable after cyclosporine therapy and unfortunately, the patient's dermatological symptoms did not respond to the therapy. Therefore, we decided to give IVIG therapy to the patient while she was taking oral methylprednisolone P.O. 4 mg per day. She received an intravenous drip infusion of human IgG at 400 mg/kg/day for 5 consecutive days in one month. This therapy was given to her five times. The lesions began to heal and no new lesions had appeared at that period. But she couldn't come to control visits after 5 IVIG infusions because of some personal reasons. 8 months later, she came with activation of the disease. There were scattered erosions around submammarian region and crusted scaly erythematous plaques with a few erosions on the scalp (Figure 1). In addition, she had erosions on the soft palate. She has been taking methylprednisolone P.O. 4 mg per day and applying topical corticosteroids on the scalp and submammarian region two times a day. Pathohistological findings at the scalp lesion revealed suprabasilar acantholysis but there was subcorneal acantholysis at the submammarian lesion specimen (Figure 2). Direct immunofluorescent technique demonstrated IgG in the epidermal layers. To characterize the autoantibodies in this case, we performed ELISA test for the detection of autoantibodies against Dsg3 and Dsg1. The values of both autoantibodies were positive. On the basis of these findings, PV and PF were diagnosed. So, we decided to increase methylprednisolone dose to 16 mg P.O. per day and give IVIG therapy to her again.

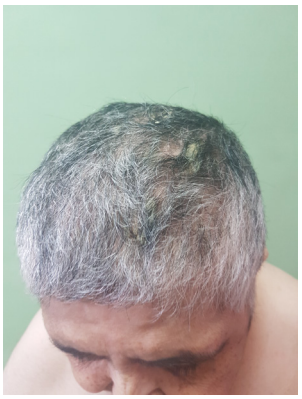

Figure 1A

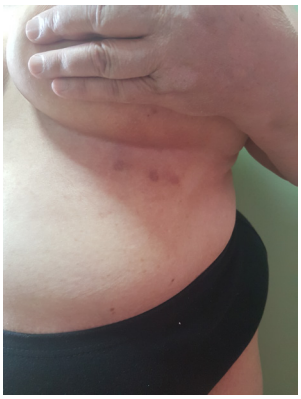

Figure 1B

Figure 1: (A) Crusted lesions on the scalp (B) Scattered erosions at the submammarian area.

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Received: June 14, 2017; **Accepted:** July 06, 2017; **Published**: July 17, 2017

Citation: Durmaz K, Dursun R, Ataseven A, Özer I, Fındik S (2017) Coexistence of Pemphigus Vulgaris and Pemphigus Foliaceus in the Same Patient at the Same Time. Dermatol Case Rep 2: 124.

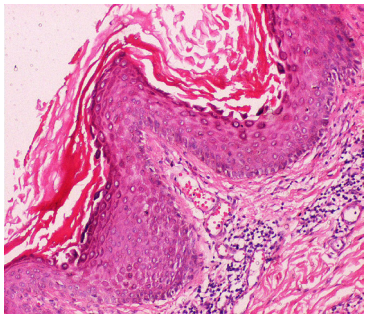

Figure 2A

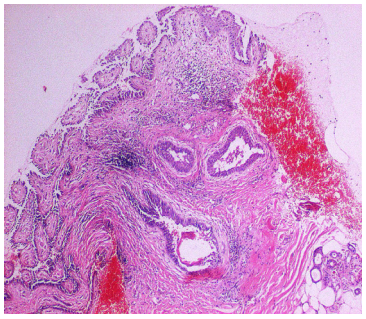

Figure 2B

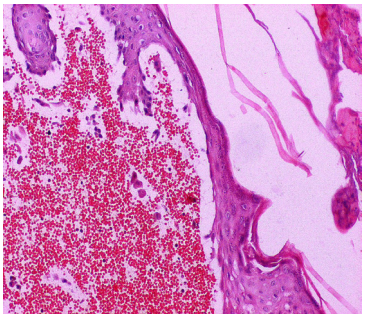

Figure 2C

Figure 2: (A): Subcornal blister with dyskeratotic acantholytic granular cells. Hematoxylin eosin (H&E) x100. (B): (x40) and (C): (x100) Intraepidermal acantholytic blister contains acantholytic cells. Dermal papillae lined by a single layer of basal keratinocytes. Hematoxylin eosin (H&E).

Discussion

The two types of pemphigus show some different features. PV is characterized by expanded cutaneous lesions with oral mucosal involvement. Blisters in PV are placed in the suprabasal layers of the epidermis. In PV, the target antigen is Dsg3 which located primarily on the oral mucosa and lower epidermis. In PF, the acantholysis exist within the upper layers of the epidermis, resulting in clinical crusts or superficial blisters without oral involvement, as the target antigen is Dsg1, which is located on the upper epidermis of the skin. Almost 60% of patients affected by PV also have circulating Dsg1 autoantibodies without any clinical symptoms of PF-like lesions [13]. The shifting between PV and PF is an uncommon situation [14]. Previous studies have suggested that qualitative changes in Dsg autoantibody profile might underlie this transition [15-17]. This transition is a reflection of qualitative and quantitative changes in the profile of developed autoantibodies against Dsg1 and Dsg3 antigens [18]. The pathogenic role of antidesmoglein in pemphigus is well known. Recent studies have demonstrated that the clinical phenotype of the disease is described by the antidesmoglein autoantibody profile and by the Dsg1 and Dsg3 tissue distribution [19].

Conclusion

In the reported cases of transformation between PV and PF, immunoblotting studies have suggested that the change in clinical features is related to a change in antibody profile [18]. Our patient displayed a mixed clinical and histopathological expression of features of both PV and PF related to co-expression of anti-Dsg3 and anti-Dsg1 antibodies. Rare cases of concurrent presence of PF and PV have been described in the literature [20,21]. So, our patient may have a rare case of pemphigus, diagnosed from clinical and histopathological findings, with detected two autoantibodies belong to PV and PF disease's pathogenesis.

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