Two Cases of Pemphigus Foliaceus with Severe Pruritus and Elevated IgE Levels

Jingfang Zhang^{1,2}, Liu Yang^{1,2}, Yamin Zhang^{1,2*}, Juan Tao^{1,2}

¹Department of Dermatology, Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430022, China

²Hubei Engineering Research Center for Skin Repair and Theranostics, Wuhan 430022, China

*Corresponding Author: Yamin Zhang, Department of Dermatology, Union Hospital, Tongji Medical College, No. 1277 Jiefang Avenue, Wuhan, Hubei, 430022, China; E-mail: 879132375@qq.com

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Abstract

We report the clinical characteristics and management of two patients with severe PF, who presented with extreme pruritus and high IgE levels. Both the two patients had common characteristics as follows

1. They were all diagnosed as PF and presented with chronic eczematous lesions
2. The patients all accompanied with intense pruritus and were resistant to diversified forms of antihistamines. Furthermore, the level of IgE antibody in their serum was significantly increased
3. Their skin lesions were accompanied with *Staphylococcus aureus* infection by secretion culture
4. After the diagnosis of PF was made, methylprednisolone (1.5 mg/kg) and antibiotics were administered, which rapidly induced remission and the levels of Dsg1 and IgE in the serum were significantly declined. The possible role and mechanism of IgE elevation in these two cases were also discussed

Keywords

Pemphigus Foliaceus; Elevated IgE; Pruritus; Chronic Eczematous Lesions; Bacterial Infection

Case Report

Pemphigus Foliaceus (PF) is a kind of autoimmune blistering diseases mediated by pathogenic autoantibodies. In contrast to pemphigus vulgaris, PF rarely invades mucosal surfaces and predominantly presents as erythema, puff pastry-like scaling and crusting. The more severe forms can present as erythroderma and these patients are prone to superinfections [1,2]. Herein, we report the clinical characteristics and management of two patients with severe PF, who presented with extreme pruritus and high IgE levels.

Patient 1 was a 55-year-old female, presenting to inpatient dermatology ward and erythrodermic on initial presentation, complained of pruritus and burning of the skin. Widespread, edematous and erythematous plaques and puff pastry-like scaling and crusting was present on her scalp, face, trunk and extremities. Yellow, greasy crust and a purulent exudate were present in the peri-orbital and peri-orificial areas. There were erosions and flaccid blisters on the extremities (Fig. 1). Patient 2 was a 71-year-old male presented with erythematous plaques, erosions, scaling and lichenified plaques, covering an extensive area of the trunk (Fig. 2). Neither the two patients had a history of allergic disease, including asthma, allergic rhinitis, parasitic infection or atopic history, nor the use of medicines which may cause elevated IgE levels, including herbal or traditional Chinese medicines during the onset and treatment of disease.

In both patients who presented with clinical pictures consistent with a chronic eczematous pathology, the diagnosis of PF was established on the basis of histopathological findings and immunofluorescence (Fig. 1). Intense pruritus was present in both cases, resistant to multiple antihistamine trials. Furthermore, the level of IgE antibody in their serum was significantly increased. Cultures from impetiginized skin lesions grew *Staphylococcus aureus*. After the diagnosis of PF was made, methylprednisolone (1.5 mg/kg) and antibiotics were administered, achieving rapid remission in both cases (Fig. 1). The serum levels of Dsg1 antigen and IgE showed a simultaneous and concomitant decline (Table 1).

In contrast to the more classic presentations of PF, our patients presented with chronic eczematous lesions, accompanied with intense pruritus. Histopathological examination revealed spongiosis, acantholysis and inflammatory cell infiltrates. These findings varied based on the location and chronicity of lesions. Therefore, in such patients, biopsies from multiple sites may be required for diagnosis [3]. The chronic eczematous lesions in these patients may have developed due to an interruption in therapy. In allergic diseases, FcεRI-dependent

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degranulation of mast cells and eosinophils of IgE are also contributors to pruritus [4]. Though both our patients presented with increased IgE levels, neither had a history of allergic diseases. While, they all developed to chronic eczematous lesions and histopathological presentation due to an interruption in therapy, indicating that both our patients may have eczema comorbidity and the elevated IgE mediated clinical symptoms of pruritus.

Besides, we observed that IgE levels were positively correlated with clinical severity in both patients, indicating that IgE may be contributors to the pathogenesis of PF. While the role and mechanism of IgE in pemphigus is still unclear, accumulating evidence suggests that increased IgE levels may play a significant role in allergic and certain autoimmune diseases, such as Systemic Lupus Erythematosus (SLE) and Bullous Pemphigoid (BP), but these elevations are not found in pemphigus [4]. In SLE, autoreactive IgE could promote disease progression by FcεRI-triggered plasmacytoid Dendritic Cells (pDCs) to secrete interferon-alpha (IFN-α).⁵ In patients with BP, besides the FcεRI-induced degranulation of mast cells and basophils, self-reactive IgE reacts with hemidesmosomal cell-surface proteins BP230 and BP180, reducing number of hemidesmosomes also contribute to the pathogenesis of BP [5]. Although the lesions in PF are induced by IgG autoantibodies directed against Dsg1, the targeted biologic therapies include rituximab (anti-CD20 monoclonal antibody) and anti-tumor necrosis factor agents have been adopted in some cases of PF [6]. We inferred that the role of IgE in PF may also related to IFN-α production or reacting with Dsg1. Therefore, the exact role and mechanism of IgE in pemphigus needs to be explored further, especially whether the elevated IgE is autoreactive antibodies.

This study has several limitations:

1. Due to the small sample size, findings may not be generalizable
2. Even though total IgE was elevated, Dsg1-specific IgE levels were not obtained. Therefore, it is difficult to elucidate whether IgE is directly involved in the pathogenesis of PF

Figure 1: The clinical characteristics and diagnostics of Patient 1. (A) Clinical manifestations before treatment. Widespread edematous erythematous plaques and puff pastry-like scaling and crusting on her scalp, face, trunk and limbs; (B) Clinical manifestations after 2 months of treatment; (C-F) The histopathological and immunopathological features of Patient 1; (C) Intraepidermal blister with Tzanck cells (Hematoxylin and Eosin stain, original magnification $\times 10$) and (D) eosinophilic spongiosis (Hematoxylin and eosin stain, original magnification $\times 20$). The deposition of (E) IgG and (F) C3 in acantholytic cells.

Figure 2: The clinical manifestations in Patient 2 before and after treatment. (A, B) Extensive erythematous plaques, erosions and scaling mainly with lichenification, mainly on the trunk. (C, D) Clinical manifestations in Patient 2 after treatment for a month.

	Patient 1	Patient 2
Age	55 years	71 years
Sex	Female	Male
Mean duration of diagnosis	6 months	3 months
Distribution of skin lesions	Widespread	Widespread
Symptoms	Severe itching	Severe itching
Fever	Yes (The highest recorded temperature was 39°C)	No
Concomitant Diseases	Erythroderma, hypoproteinemia, mild anemia, pelvic and pleural effusion	Eczema, small intestinal stromal tumor, mild anemia, kidney calculi and hepatic cyst
Histopathological diagnosis	pemphigus foliaceus	pemphigus foliaceus
Nikolsky sign	positive	positive
Immunofluorescence	IgG and C3	IgG
Autoantigen	Dsg1	Dsg1
Before treatment	189.1 U/ML	174.2 U/ML
After treatment	137.6 U/ML	102.8 U/ML
Total serum IgE (normal value)	Increased	Increased
Before treatment	1-190 IU/ml	1-190 IU/ml
After treatment	851 IU/ml	1078 IU/ml
	145 IU/ml	621.5 IU/ml
Cultures (skin)	<i>Staphylococcus aureus</i>	<i>Staphylococcus aureus</i>
Treatment	Methylprednisolone, 40 mg/day; Fodum fusidate, 1g/day; IVIG 5g/day	Methylprednisolone, 60 mg/day; Cefdinir 300 mg/day
Medication withdraw	Yes	Yes
Clinical outcome	Stable and discharged	Stable and discharged

Table 1: Clinical, histopathological and immunopathological features of Patients 1 and 2.

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Conclusion

In conclusion, our cases with clinical features and symptoms of chronic eczema and increased IgE levels, highlight the importance of considering PF in the differential diagnosis of eczematous diseases, especially in those with long-standing disease. Furthermore, our study suggests that the role of IgE in the PF pathogenesis needs to be explored further.

Conflict of Interest

There is no conflict of interest to declare.

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The 2 patients in the manuscript have given written informed consent for publication of their case details.

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