Case Report

A Case of Psoriasis and Pemphigus Foliaceous in a 55-year-old Filipino

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Abstract

Pemphigus foliaceous is a rare autoimmune blistering disease, while psoriasis is a common immune-mediated inflammatory skin disease. The coexistence of psoriasis and pemphigus foliaceous has rarely been reported. We report a case of a 55-year-old Filipino female with an 8-year history of chronic plaque-type psoriasis biopsy-proven. After 5 years, she developed generalized flaccid bullae and crusted erosions over the face, trunk, and extremities, with no mucous membrane involvement. Skin punch biopsy, direct immunofluorescence, and enzyme-linked immunosorbent assay were consistent with pemphigus foliaceous. The combination of topical corticosteroids and oral methotrexate was selected as the therapeutic approach, leading to a notable improvement in the patient's condition. This case report underscores the significance of identifying the simultaneous presence of psoriasis alongside autoimmune blistering diseases like pemphigus foliaceous. Examining predisposing and triggering factors, performing re-biopsy, and further work-up as the disease evolves may yield more profound insights. Nonetheless, effectively managing this condition poses a significant challenge.

Keywords: Desmoglein-1, direct immunofluorescence, methotrexate, pemphigus foliaceous, psoriasis

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Submitted: 24-01-2024, **Revised:** 18-04-2024, **Accepted:** 22-04-2024, **Published:** 31-05-2024.

Introduction

Pemphigus foliaceous is a rare autoimmune blistering disease that presents with flaccid blisters on the skin with an annual incidence of 0.8/million.[1] Psoriasis is another immune-mediated dermatological condition that affects 0.1%–3% of the population. It characterizes inflammatory scaly plaques and poses an increased risk of developing destructive arthritis.[2] It has been reported to be linked with different autoimmune diseases, with bullous pemphigoid as the most common blistering disorder associated with it.[3] In this case report, pemphigus foliaceous was found to be coexistent with psoriasis. The coexistence of psoriasis and pemphigus foliaceous is rarely seen up to this day. The administration of methotrexate combined with topical corticosteroids showed improvement in this case of psoriasis and pemphigus foliaceous.

Access this article online	
Quick Response Code:	Website: https://journals.lww.com/jpds
	DOI: 10.4103/JPDS.JPDS_8_24

Case Report

The patient is a 55-year-old Filipino female with an 8-year history of relapsing chronic plaque-type psoriasis. She was managed with methotrexate 7.5 mg/week during flare-ups. She presented with a sudden eruption of flaccid vesicles

How to cite this article: Ollet AM, Garcia MD, Mendoza CG. A case of psoriasis and pemphigus foliaceous in a 55-year-old Filipino. J Philipp Dermatol Soc 2024;33:21-4.

and crusted erosions over the face, trunk, and upper and lower extremities [Figure 1]. Lesions were associated with burning pain but with no fever.

During the initial consultation, a dermatological examination revealed flaccid vesicles, bullae, and indurated plaques, with some erosions over the face, trunk, and upper and lower extremities. In addition, multiple well-defined, irregularly shaped erythematous plaques with silvery white scales were also present on the scalp, ears, and postauricular area. No oral or genital lesions were seen.

The patient's previous biopsy results from 2016 showed mounds of parakeratosis with neutrophils, acanthosis in the epidermis, and superficial perivascular lymphocytic infiltrates with few eosinophils seen in the dermis, consistent with a final diagnosis of psoriasis vulgaris. At the time of consultation, a 4-mm skin punch biopsy was taken from a fresh bulla for routine histologic examination, and another specimen was taken on perilesional skin for direct immunofluorescence (DIF). The histopathologic examination using hematoxylin and eosin stain revealed a subcorneal split with acantholytic keratinocytes in the granular layer. DIF demonstrated intercellular deposition of complement (C3) and immunoglobulin G (IgG) grade +2 in the epidermis [Figure 2]. Euroimmun enzyme-linked immunosorbent assay (ELISA) for desmoglein 1 was positive with a ratio of 7.616, while desmoglein 3 was negative with a ratio of 0.173. Other diagnostic tests such as complete blood count, urinalysis, fasting blood sugar, lipid profile, and kidney and liver function tests were unremarkable.

With the history, clinical, histopathologic, immunofluorescence, and ELISA findings, a diagnosis of psoriasis and pemphigus foliaceous was made. The patient was started on methotrexate 15 mg weekly for 3 weeks. Antihistamines (loratadine) were also given. Regarding wound management, mild soap, normal saline solution compress, and halobetasol propionate 0.05% ointment 30 g combined with petroleum jelly 50 g were advised. There was remarkable improvement within the initial 2 weeks of treatment, manifesting as drying and reduction in lesion size, as well as the absence of new bullae formation. The patient continued to demonstrate significant and sustained progress with the 15 mg methotrexate dose in the 2nd month of treatment, which was seen as a decrease in body surface area, erythema, and erosions [Figure 3].

Discussion

Pemphigus foliaceous is a rare autoimmune blistering disease that produces IgG autoantibodies against an intercellular adhesion glycoprotein seen in the stratum granulosum of the epidermis, which is desmoglein-1. Binding these antibodies to desmoglein-1 would lead to loss of intercellular adhesion of the keratinocytes, which leads to

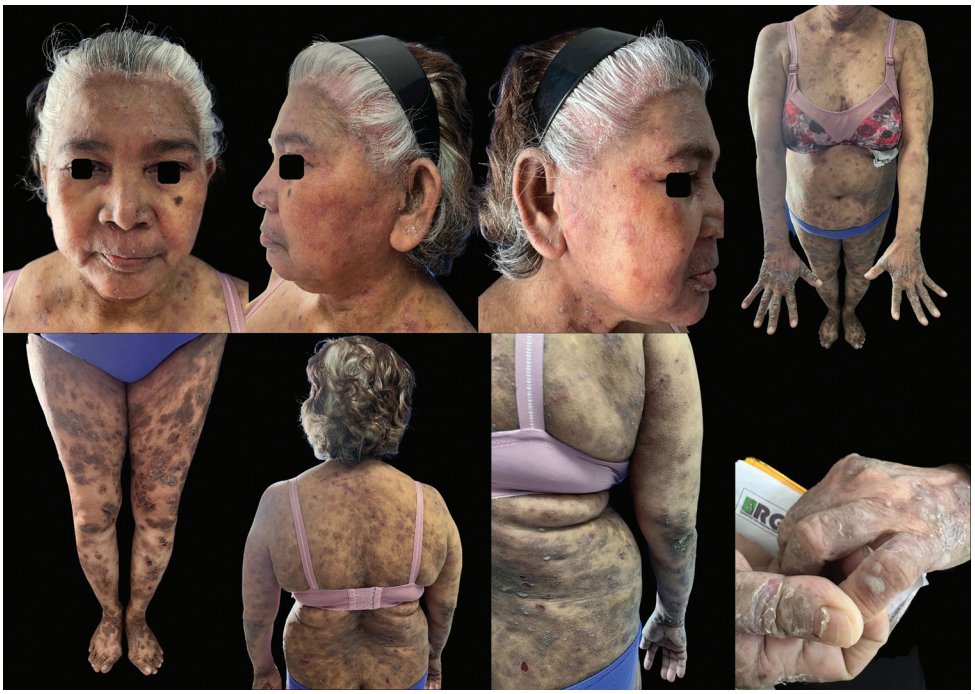

Figure 1: Initial consult. Physical examination findings of a 55-year-old female with psoriasis and pemphigus foliaceous. The lesions are described as multiple, well-defined irregularly shaped erythematous plaques with thick white scales on the scalp, ears some on the trunk and extremities as well as multiple, flaccid vesicles and crusted erosions on the face, trunk and extremities

the formation of superficial flaccid blisters and bullae that easily rupture, leading to erosions.[4] Meanwhile, psoriasis is a T-cell-mediated chronic inflammatory disorder with an abnormal cytokine and chemokine profile that causes excessive keratinocyte proliferation.[2] In a retrospective study of seven patients with psoriasis, four out of seven eventually developed pemphigus foliaceous with a range of onset from 4 to 30 years.[5] In our case, the onset was eight years before developing pemphigus foliaceous. A much larger retrospective cohort study found that patients with psoriasis have more than three times increased risk of developing pemphigus compared to the general population. The risk is higher in female patients with psoriasis.[6]

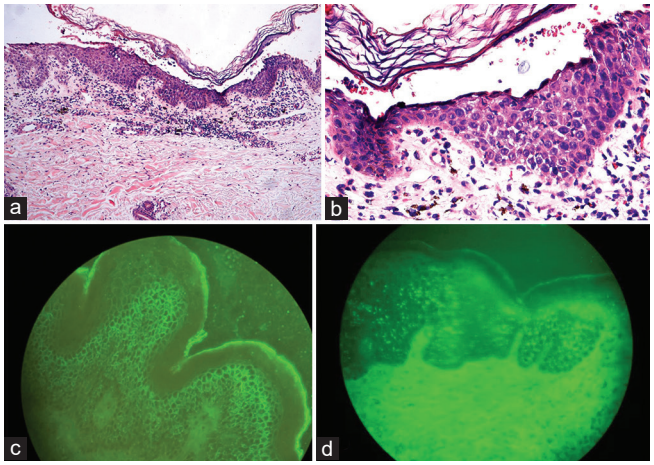

Figure 2: (a) Subcorneal split and spongiotic epidermis (H and E, ×10). (b) Acantholytic keratinocytes in the granular layer (H and E, ×40). (c) Direct immunofluorescence studies showed (+) intercellular deposition of immunoglobulin G, grade 2. (d) and (+) intercellular deposition of C3, grade 2

While it is theoretically possible for a person to have both conditions, it would be an unusual case.[7] Different theories have been proposed regarding the co-occurrence of both diseases. Psoriatic lesions have high levels of tissue plasminogen activator, which was proposed to be responsible for the acantholysis in pemphigus.[8] Suppression of T-lymphocyte function in psoriasis may lead to overactivity of the humoral immune system, producing auto-antibodies.[3,5,8] Genetic predisposition may also play a part in the pathogenesis. HLA-DRB1 alleles have been mentioned in many studies in both psoriasis and pemphigus foliaceous, suggesting a particular linkage to both diseases.[8]

Clinical manifestations of psoriasis commonly present as a well-demarcated raised plaque with a white scaly surface. Meanwhile, pemphigus foliaceous primary lesions are small flaccid blisters that are typically not found. It presents as scaly, crusted erosions on an erythematous base. Histopathology findings of pemphigus foliaceous would show acantholysis below the stratum corneum and in the granular layer. The diagnostic hallmark of pemphigus foliaceous is a positive DIF study on normal-looking perilesional skin, which shows the presence of IgG autoantibodies and C3 in the epidermis.[2] In this case,

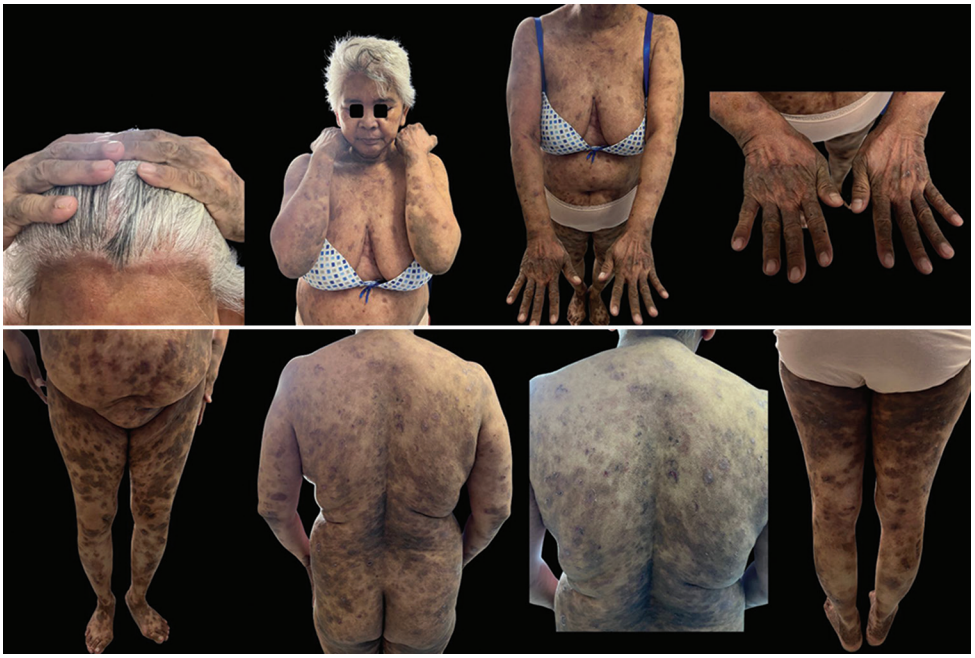

Figure 3: Physical examination findings after 2nd month of consult. There is noted decrease in body surface area, erythema and erosions. The lesions are described as multiple, well-defined irregularly shaped erythematous plaques with white scales on the scalp as well as multiple, well-defined, irregularly shaped hyperpigmented patches and plaques some with erosions on the trunk and extremities

acantholysis occurred between the stratum corneum, granular layer, and subcorneal split. Furthermore, the intercellular deposition of IgG and C3 in the epidermis showed a grade of +2, consistent with pemphigus foliaceous. Antigen-specific ELISA has proven to be a highly sensitive and specific tool in diagnosing pemphigus. In the case at hand, desmoglein-1 yielded a positive result with a ratio of 7.616, whereas desmoglein-3 returned a ratio of 0.713, further substantiating the diagnosis of pemphigus foliaceous. The first-line treatment for pemphigus foliaceous is oral corticosteroids. However, this case also presented with concomitant psoriasis vulgaris, in which oral corticosteroids were contraindicated; hence, topical steroids and methotrexate were used.[2] The coexistence of both diseases poses an arduous dilemma for their management.

Methotrexate has diverse clinical benefits because of its anti-inflammatory, cytotoxic, and immunological modulatory properties.[9] It is a dihydrofolate reductase inhibitor that is believed to act directly on inhibiting epidermal hyperproliferation and also has immunosuppressive effects.[2] In a case report done by Tripathy *et al*., methotrexate and mycophenolate mofetil were used to treat a patient with co-occurrence of psoriasis and pemphigus foliaceous. Due to the incomplete resolution of blisters, oral corticosteroids such as prednisolone were added, which showed continuous resolution of lesions and no recurrence of flare-ups on discontinuing prednisolone.[8] In another case report done by Zheng *et al*., methotrexate combined with methylprednisolone was used on a patient with psoriasis and pemphigus vulgaris, which resulted in gradual resolution of lesions.[10]

Patient education on long-term monitoring and management is vital for patients with this type of disease. Measuring tools to assess the severity and response to treatment in this case, such as psoriasis area and severity index and pemphigus disease area index is quite challenging due to disease overlap. ELISA titers should be done every 3–6 months to monitor pemphigus disease activity while undergoing management. Performing a re-biopsy and further work-up should the disease evolve is valuable. Furthermore, a comprehensive review and plan of additional treatments may be warranted in this case in the situation of possible relapse. This may provide more vital information for possible standardized therapy in the future.

Conclusion

This case report underscores the significance of identifying the simultaneous presence of psoriasis alongside autoimmune blistering diseases like pemphigus foliaceous. Examining predisposing and triggering factors, performing re-biopsy, and further work-up as the disease evolves may yield more profound insights. Nonetheless, effectively managing this condition poses a significant challenge. Thus, a therapeutic pathway for managing this condition is vital.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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