Case Report: An Unusual Presentation of Pemphigus Foliaceus

Zahra Albalooshi[1], Mutia Al Jabi[2] and Ahmed Aboeldahab[3]*

[1]Family Medicine Department, Mediclinic Hospital, Abu Dhabi, United Arab Emirates

[2]Basic Medical Sciences, College of Medicine, RAK university, RAK, United Arab Emirates

[3]Basic Medical Sciences, College of Medicine, University of Sharjah, Sharjah 27272, United Arab Emirates

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*Corresponding author: aboeldahaba32@yahoo.com

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Abstract

Pemphigus foliaceus (PF) is a rare autoimmune blistering disease that specifically targets desmoglein-1, leading to superficial skin erosions and significantly impairing patients' quality of life.[1] The coexistence of multiple autoimmune conditions in a single patient raises important questions regarding potential shared pathophysiological mechanisms that may underlie autoimmune disease clustering. This case report presents a 59-year-old female who was recently diagnosed with PF, with a medical history notable for type 2 diabetes, hypothyroidism, and a past diagnosis of pituitary adenoma. She initially presented with worsening skin lesions, which were confirmed as PF through immunofluorescence and histological analysis. Given the presence of multiple autoimmune conditions, this case highlights the possibility of PF being linked to an increased susceptibility to other immune-mediated diseases, particularly metabolic disorders such as diabetes. The management of PF in this patient posed significant challenges due to the necessity of balancing immunosuppressive therapy with the need to control her comorbidities. Immunosuppressive treatment, while essential for PF management, required careful monitoring to prevent exacerbation of her diabetes and thyroid dysfunction. This underscores the importance of a personalized, multidisciplinary approach that integrates dermatology, endocrinology, and internal medicine expertise.[2,3] By examining this case, we aim to emphasize the need for further research into the interplay between PF and systemic autoimmune diseases. Moreover, this report reinforces the importance of screening PF patients for concurrent autoimmune conditions to facilitate early detection, optimize management strategies, and improve long-term patient outcomes.

Keywords: Pemphigus foliaceus, Type 2 diabetes, Adenoma, Autoimmune, Hypothyroidism

Introduction

Pemphigus foliaceus (PF) is an autoimmune blistering condition marked by the generation of autoantibodies targeting desmoglein-1, an essential element of desmosomes that facilitate adhesion among keratinocytes.[1] This disorder results in the development of superficial, fragile blisters and erosions predominantly impacting the skin, especially in seborrheic regions like the head, face, and chest. PF can severely diminish quality of life due to pain, discomfort, and the potential for subsequent infections, while also presenting considerable treatment challenges. Management frequently involves the administration of systemic immunosuppressive medications, including mycophenolate mofetil (MMF) and prednisone, aimed to diminish the autoimmune response and regulate disease activity.[4,5] Nonetheless, these drugs may have considerable adverse effects and could complicate the care of patients with pre-existing diseases, including diabetes mellitus. This case report examines a 59-year-old female patient with a history of diabetes diagnosed with pemphigus foliaceus. The connection between her underlying diabetes and the immunosuppressive agents—MMF and prednisone—presents a complicated clinical situation. We intend to examine the implications of her treatment protocol, the management approaches utilized for her autoimmune disease and

diabetes, and the clinical outcomes noted in this particular case. This patient's path illustrates the complexities of managing autoimmune disorders alongside comorbidities.

A 59-year-old female patient, with a history of pemphigus foliaceus, has been sent from dermatology to the family medicine department for a review of her CBC and metabolic profile, as she is now on immunosuppressants.

Cushingoid appearance characterized by generalized crusted papules and plaques located on the face, scalp, chest, and back. The patient exhibited an absence of mucosal lesions and nail changes. The patient reported recurrent joint pain primarily localized in the knees.

Assessment involved the collection of blood samples for comprehensive laboratory analysis, which included serological testing for specific antibodies. A skin biopsy was conducted to assess histopathological changes revealing interface dermatitis, a characteristic change in pemphigus foliaceus, which was further evaluated using direct immunofluorescence. The diagnosis of pemphigus was confirmed through direct immunofluorescence, which revealed a sub-corneal pustule consistent with pemphigus foliaceus, along with positive IgG and C3 on direct immunofluorescence.

Following the confirmation of P.F diagnosis, the patient was initiated on prednisolone, Mycophenolate Mofetil, and pimecrolimus. Considering the patient's history of diabetes mellitus, Dapagliflozin-metformin was resumed and the patient was instructed to monitor closely and schedule follow-ups to address any potential disease or drug-related adverse reactions. Further information about the medications given will be found in table 1.

Table 1: Patient’s medications and doses for PF and diabetes.

Medication	Dose
Prednisolone	5 mg
Mycophenolate Mofetil	500 mg
Pimecrolimus Cream	1%
Dapagliflozin-Metformin	5 mg - 1000 mg

The patient consistently attended appointments with the family medicine department, where routine blood work and urinalysis were conducted to monitor overall health, assess diabetes management, and evaluate the impact of medications for pemphigus foliaceus. After laboratory evaluation, the patient exhibited abnormal findings, suggesting uncontrolled diabetes potentially worsened by steroid therapy, despite continuous metformin treatment. Hematological analysis indicated an increase in white blood cell (WBC) count, red blood cell (RBC) count, immature granulocytes, and neutrophils, while mean corpuscular hemoglobin (MCH), mean corpuscular hemoglobin concentration (MCHC), and mean corpuscular volume (MCV) were decreased. The patient was referred to orthopedics for additional evaluation due to ongoing knee pain. An MRI demonstrated multifocal bilateral intramedullary lesions indicative of steroid-induced osteonecrosis, alongside an area of abnormal enhancement in the left upper tibial diaphysis, linked to a sinus tract and external fluid collection, suggesting a potential underlying infection.

Discussion

Pemphigus foliaceus (PF) is a rare, chronic autoimmune blistering illness that mostly affects the skin, resulting in superficial blisters caused by acantholysis within the epidermis. Unlike pemphigus vulgaris, PF is often limited to the upper layers of the epidermis and frequently spares the mucous membranes. IgG antibodies targeting desmoglein 1, a desmosomal protein required for keratinocyte adhesion, are the most common mediators of the syndrome. This case report focuses on numerous unique elements of PF, providing useful insights into its clinical presentation, management problems, and therapeutic response.

Because of the variety in PF presentations, particularly in non-endemic areas, the diagnosis may be delayed or misdiagnosed for other dermatoses such as seborrheic dermatitis, eczema, or psoriasis. A biopsy verified acantholysis in the top epidermis, and direct immunofluorescence revealed intercellular IgG and C3 deposits compatible with PF. These findings highlight the significance of histology and immunofluorescence in identifying PF from comparable diseases.

PF management can be complicated by its chronic nature and the potential side effects of long-term immunosuppressive therapy. Corticosteroids are still the first-line treatment; however, adjuvant medications such as azathioprine or mycophenolate mofetil are commonly used to minimize steroid dependence.[4] Newer biologic treatments, such as rituximab, show promise in refractory patients, providing targeted immunomodulation by depleting B-cells and lowering antibody synthesis. A monoclonal antibody called rituximab, which targets CD20+ B cells, has shown promise in treating moderate-to-severe or refractory PF. Patients who experience severe steroid-related problems, such as uncontrolled diabetes, hypertension, or osteoporosis, or who are unable to treat their condition with corticosteroids and conventional immunosuppressants, are usually given this option. Patients who experience frequent relapses or a disease that progresses quickly may also benefit from it early.[5,6] Rituximab is a relevant second-line option because of the patient's history of diabetes and variable hematological parameters, which emphasize the significance of reducing systemic corticosteroid exposure. During rituximab therapy, close observation is necessary for infusion reactions, hepatitis B reactivation, and neutropenia.

This example highlights the risk of problems associated with PF, including secondary infections caused by decreased skin integrity and adverse effects from extended immunosuppression. Patients with PF frequently have an extended disease history, and long-term care necessitates striking a balance between treating symptoms and minimizing treatment-related morbidity. The prognosis is often variable, with illness severity determined by both genetic predisposition and environmental factors. The patient had high neutrophil and WBC counts, which may indicate a subclinical infection, underlying inflammation, or corticosteroid-induced leukocytosis, a known side effect of systemic steroid therapy. A reactive marrow response, possibly brought on by stress or immunosuppression, is further suggested by the rise in immature granulocytes. Furthermore, a higher RBC count along with lower MCV, MCH, and MCHC could indicate hemoconcentration from dehydration, early or concealed microcytic anemia, or even iron-deficiency anemia. Since diabetic patients are more susceptible to infection, volume changes, and metabolic problems, these hematologic abnormalities should be cautiously watched in PF patients undergoing systemic therapy.

This paper adds to the growing body of evidence on PF, particularly in terms of differential diagnosis and tailored treatment regimens. As new medicines become available, case studies like this can help clinicians provide effective, patient-centered care for rare autoimmune illnesses such as PF. Additionally, this case suggests that PF and other autoimmune disorders may share pathophysiological pathways. Multiple autoimmune diseases, including type 1 diabetes and hypothyroidism, coexisting with PF may indicate a more widespread immune dysregulation that may be connected to abnormal T-cell regulation or variations in the HLA gene. When PF patients exhibit new or unexplained symptoms, autoimmune clustering—a well-known phenomenon—indicates the necessity of systemic screening.[7] More research is needed to optimize treatment regimens, eliminate side effects, and improve the quality of life for those affected by PF.

Conclusion

This case highlights the complex issues surrounding the diagnosis and treatment of the uncommon autoimmune blistering disease pemphigus foliaceus. A multidisciplinary approach that stresses both successful disease control and the prevention of treatment-related consequences is necessary given the patient's concurrent comorbidities, which include diabetes, hypothyroidism, and a history of pituitary adenoma.[3] The diagnosis was confirmed with the help of immunofluorescence and histology, which demonstrated their vital roles in distinguishing PF from other dermatoses. The possible overlap in pathophysiological pathways between autoimmune disorders is also highlighted in this paper, underscoring the significance of thorough screening for other autoimmune conditions in patients with PF. Clinicians can better predict problems and customize treatment strategies to meet the needs of each patient by implementing such approaches. It encourages more investigation into the common processes behind autoimmune diseases and the creation of focused treatments to enhance patient outcomes and quality of life.

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