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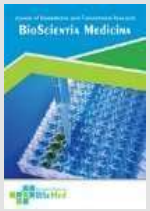

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An Unusual Case of Pemphigus Foliaceus Arising in a Patient with Psoriasis Vulgaris

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A B S T R A C T

Background: Pemphigus foliaceus (PF) is a rare autoimmune blistering disease characterized by superficial, fragile blisters. Psoriasis vulgaris, a chronic inflammatory skin condition, has been rarely associated with PF. This case report presents an unusual instance of PF developing in a patient with a history of psoriasis vulgaris. **Case presentation:** A 54-year-old Indonesian woman presented with a one-year history of scaly skin and reddish spots across her body, worsening over the past week. She had a prior diagnosis of psoriasis vulgaris and was undergoing methotrexate therapy (15 mg/week) without significant improvement. Three months prior, she developed loose blisters on her back that spread to her hands, rupturing easily and leaving painful sores. The patient denied any mucosal involvement. One week before her presentation, her symptoms worsened following relocation-related stress. Dermatological examination revealed generalized multiple erythematous patches with scales, some with ruptured blisters leaving erosions, and a positive Nikolsky sign. Histopathological examination confirmed PF. The patient was treated with intravenous methylprednisolone, oral erythromycin and paracetamol, topical mupirocin, and clobetasol. After one month, due to a lack of improvement, azathioprine was added, leading to lesion improvement without side effects. **Conclusion:** This case highlights the rare but potential development of PF in patients with psoriasis vulgaris. The complex interplay between these two conditions warrants further investigation. Early diagnosis and appropriate treatment are crucial for managing PF and improving patient outcomes.

1. Introduction

Pemphigus foliaceus (PF) stands as a rare, chronic autoimmune blistering disease that primarily targets the skin. Its distinctive characteristic is the disruption of cell-cell adhesion within the epidermis, the outermost layer of the skin, leading to the formation of superficial, fragile blisters. This disruption is triggered by the presence of autoantibodies specifically directed against desmoglein 1 (Dsg1), a transmembrane glycoprotein that plays a critical role in maintaining the integrity of the epidermis. These autoantibodies interfere with Dsg1's adhesive function, resulting in a loss of cell cohesion and the subsequent formation of blisters. Typically, PF manifests with small, flaccid blisters that are prone to easy rupture, leaving behind painful erosions and crusts. These lesions often occur extensively and can potentially affect any part of the body, although they commonly appear on the face, scalp, chest, and back. A key distinguishing feature of PF is that it rarely affects the mucous membranes, unlike other forms of pemphigus, such as pemphigus vulgaris.[1-4]

The precise cause of PF remains elusive, but current understanding suggests it involves a complex interplay of genetic and environmental factors. Research has identified certain genetic predispositions, and it is believed that environmental triggers, such as medications, infections, and ultraviolet radiation, may contribute to the development of the disease. In contrast, psoriasis

vulgaris is a chronic inflammatory skin disease characterized by well-defined, erythematous plaques with silvery scales. It is a relatively common condition, estimated to affect approximately 2-3% of the general population. The underlying mechanism of psoriasis involves an abnormal immune response, which leads to increased proliferation of skin cells and inflammation.[5-7]

The association between PF and psoriasis vulgaris is rare and not well understood. Some studies have suggested a possible link, with PF developing in patients with a history of psoriasis. However, the exact mechanisms underlying this association remain unclear.[8-10] This case report describes an unusual presentation of PF in a patient with a history of psoriasis vulgaris. The patient's clinical presentation, histopathological findings, and treatment course are discussed, highlighting the challenges in managing this complex condition.

2. Case Presentation

A 54-year-old Indonesian woman presented to the emergency department with a chief complaint of scaly skin and reddish spots covering most of her body. These symptoms had progressively worsened over the preceding week, causing significant distress. The patient recounted that her skin concerns began approximately one year prior, initially manifesting as localized patches of dryness and redness. However, over the past week, these patches had become more widespread and pronounced, accompanied by the development of new, concerning lesions. Three months prior to her presentation, the patient noticed the emergence of loose, fluid-filled blisters, primarily localized to her back. These blisters gradually spread to her hands and arms, causing discomfort and worry. The blisters were fragile and prone to rupture, leaving behind painful sores that often exuded an unpleasant odor. The patient emphasized that the skin lesions were not associated with any itching or fever. Additionally, she denied any mucosal involvement, such as lesions in her mouth or genital region. Seeking relief, the patient had previously visited a local clinic, where she was diagnosed with an allergic reaction. She received a course of allergy medications and antibiotics, but unfortunately, her condition did not improve. The patient could not recall the specific names of the medications she had received. One week prior to her presentation at the emergency department, the patient's symptoms dramatically escalated. The previously localized patches of dryness and redness expanded, coalescing to cover a larger surface area of her body. New blisters continued to form and rupture, leading to the development of additional painful sores. The patient noted that this exacerbation of her symptoms coincided with her recent relocation from Kalimantan to Solo. She attributed the worsening of her skin condition to the stress and emotional upheaval associated with the move, particularly the separation from her husband and child. The patient had a significant past medical history. One year prior, she was diagnosed with psoriasis vulgaris, a chronic inflammatory skin condition. She had been undergoing treatment with methotrexate, an immunosuppressive medication, at a dose of 15 mg per week. However, despite consistent adherence to the medication regimen, her skin condition had not shown any substantial improvement. In addition to psoriasis vulgaris, the patient also reported a history of diabetes mellitus and hypertension. However, she admitted to not taking her prescribed medications regularly for these conditions. The patient denied any history of similar skin complaints, malignancy, or atopy. The patient's family history was unremarkable for similar skin conditions or malignancy. The patient was a housewife who had recently relocated from Kalimantan to Solo. On physical examination, the patient appeared moderately ill. Her vital signs were as follows: blood pressure 141/82 mmHg, pulse rate 129 beats per minute, respiratory rate 20 breaths per minute, and temperature 37.3°C. Her pain level, as assessed by the visual analog scale (VAS), was 4 out of 10. The patient's nutritional status was normoweight, with a body mass index (BMI) of 23.3 kg/m². There was no palpable enlargement of lymph nodes. The

dermatological examination revealed striking abnormalities. The patient's skin exhibited generalized multiple erythematous patches, some of which were confluent. These patches were covered with scales, and some areas showed ruptured blisters with underlying erosions. The Nikolsky sign, a clinical test used to assess skin fragility, was positive. The Pemphigus Disease Area Index (PDAI) score, a tool used to quantify the severity of pemphigus, was 48, indicating extensive severe disease. The patient's scalp also appeared red and scaly. However, there were no abnormalities observed in the mucosa or nails (Table 1).

To further investigate the patient's condition, a skin biopsy was obtained and sent for histopathological examination. The biopsy specimen was processed and stained with hematoxylin and eosin (H&E) for microscopic evaluation. The epidermis, the outermost layer of the skin, showed parakeratosis, which is characterized by the retention of nuclei in the stratum corneum, the outermost layer of the epidermis. Acantholytic areas were also observed, indicating a loss of cell-to-cell adhesion between keratinocytes, the predominant cell type in the epidermis. Additionally, there were subcorneal clefts, which are splits or separations between the stratum corneum and the underlying layers of the epidermis. Infiltration of myoepithelial cells and eosinophils, both types of inflammatory cells, was also noted within the epidermis. The dermis, the layer of skin beneath the epidermis, showed infiltration of lymphocytes, another type of inflammatory cell. However, there were no signs of malignancy, such as abnormal cell growth or atypical cellular features. In addition to the H&E staining, direct immunofluorescence (DIF) was performed on the skin biopsy specimen. DIF is a technique used to detect the presence and deposition of autoantibodies, which are antibodies that mistakenly target the body's own tissues, within the skin. Positive IgG deposition was observed in the subcorneal layer of the epidermis. This finding indicates the presence of autoantibodies of the IgG class, which are commonly found in autoimmune blistering diseases like PF. Taking into account the patient's clinical presentation, which included the development of fragile blisters and erosions on her skin, along with the histopathological findings of acantholysis, subcorneal clefts, and inflammatory cell infiltration, and the positive IgG deposition on DIF, a diagnosis of pemphigus foliaceus (PF) was established (Table 2).

Given the patient's diagnosis of PF and its severity, a comprehensive treatment plan was initiated. The patient was advised to follow a high-protein diet consisting of 1700 kcal per day, with an emphasis on consuming extra egg whites. This dietary modification aims to support the body's healing process and maintain adequate nutritional status. Intravenous (IV) methylprednisolone was administered at a dose of 62.5 mg every 24 hours. Methylprednisolone is a corticosteroid medication that helps reduce inflammation and suppress the immune system, which is crucial in managing autoimmune diseases like PF. Oral erythromycin was prescribed at a dose of 500 mg three times a day. Erythromycin is an antibiotic that also has anti-inflammatory properties and can be helpful in treating skin infections that may occur secondary to PF lesions. Oral paracetamol was provided at a dose of 500 mg every 8 hours as needed for pain relief. Paracetamol is a common over-the-counter pain reliever that helps manage the discomfort associated with PF lesions. Topical mupirocin 2% ointment was advised to be applied twice daily to any areas of erosion or broken skin. Mupirocin is a topical antibiotic that helps prevent and treat bacterial infections in skin lesions. Topical clobetasol 0.05% ointment, mixed with petroleum jelly in a 2:1 ratio, was prescribed for application twice daily to the entire body. Clobetasol is a potent topical corticosteroid that helps reduce inflammation and itching associated with PF. The patient was also referred for a consultation with the endocrinology department to optimize the management of her diabetes mellitus and hypertension. Proper management of these comorbidities is essential for overall health and may indirectly improve the

treatment response of PF. After one month of this initial treatment regimen, the patient returned to the outpatient clinic for a follow-up evaluation. She reported some improvement in her condition, noting that the reddish patches on her skin had lessened in intensity. However, she still expressed concerns about the persistence of scales in some areas. In response to the patient's feedback and the clinical evaluation, her treatment plan was adjusted. Oral azathioprine was added to her medication regimen at a dose of 50 mg twice daily. Azathioprine is an immunosuppressant medication that can help control the autoimmune response in PF and reduce the need for high-dose corticosteroids. Oral cetirizine was also introduced at a dose of 10 mg once daily. Cetirizine is an antihistamine that can help relieve itching, which may be present in some cases of PF. The topical corticosteroid was changed to desoximetasone cream 0.025%, mixed with petroleum jelly in a 1:2 ratio, and applied twice daily. This adjustment aimed to provide continued anti-inflammatory effects while potentially minimizing the risk of skin thinning associated with long-term use of potent corticosteroids like clobetasol. The patient was encouraged to frequently apply pseudoceramide cream to her skin. Pseudoceramide is a type of moisturizer that can help improve skin hydration and barrier function, which can be compromised in PF. After two weeks of following this modified treatment plan, including the addition of azathioprine, the patient showed further improvement in her skin lesions. The remaining scales diminished, and no new blister formation was observed. Importantly, the patient did not experience any adverse side effects from the medications (Table 3).

Table 1. Anamnesis, physical examination, and dermatological examination.

Category	Details
Anamnesis	**Chief complaint: Scaly skin and reddish spots all over the body, worsening over the past week.**
History of present illness: 3 months prior: Loose blisters appeared on the back and spread to both hands.	
Blisters ruptured easily, causing painful sores with an unpleasant odor.	
No fever, itching, or wounds in the eyes, mouth, or genitals.	
Previous treatment at a clinic with allergy medication and antibiotics, but no improvement.	
1 week prior: Symptoms worsened, with more wounds and dry, scaly skin spreading to almost the entire body.	
Symptoms began after the patient moved from Kalimantan to Solo.	
The patient attributes worsening symptoms to stress from being separated from her husband and child.	
Past medical history: Diagnosed with psoriasis vulgaris 1 year prior.	
Rountine methotrexate 15mg/week PO tablets.	
History of diabetes mellitus and hypertension, but not routinely taking medication.	
No history of similar complaints, malignancy, or atopy.	
Family history: No family history of similar skin conditions.	
No family history of malignancy.	
Social history: Housewife.	
Recently moved from Kalimantan to Solo.	
Physical Examination	**General appearance: Moderately ill.**
Vital signs: Blood pressure: 141/82 mmHg	
Pulse rate: 129 beats/minute	
Respiratory rate: 20 breaths/minute	
Temperature: 37.3°C	
Visual Analog Scale (VAS): 4	
Nutritional status: Normoweight with a body mass index (BMI) of 23.3 kg/m²	
Lymph nodes: No enlargement.	
Dermatological Examination	**Skin: Generalized multiple erythematous patches, some confluent.**
Scales present on the patches.	
Ruptured blisters with erosions in some areas.	
Positive Nikolsky sign.	
PDAI score: 48 (extensive severe)	
Scalp: Red and scaly.	
Mucosa and nails: No abnormalities.	

Table 2. Histopathological examination and diagnosis.

Category	Details
Histopathological examination	**Epidermis: Parakeratosis, acantholysis, subcorneal clefts, myoepithelial and eosinophil infiltration.**
Dermis: Lymphocyte infiltration, no signs of malignancy.	
Direct Immunofluorescence: Positive IgG deposition in the subcorneal layer.	
Diagnosis	Pemphigus foliaceus.

Table 3. Treatment and follow-up.

Category	Details
Initial treatment	**Diet: High-protein diet (1700kcal) with extra egg white.**
Medications:	
- Methylprednisolone 62.5mg/24 hours IV.	
- Erythromycin 3x500mg tablets by mouth.	
- Paracetamol 500mg/8 hours tablets by mouth.	
- Mupirocin 2% ointment applied twice daily to erosions.	
- Clobetasol 0.05% ointment mixed with petroleum jelly (2:1) applied twice daily to the entire body.	
Other: Referral for consultation with the endocrinology department.	
Follow-up (1 month)	**Patient complaints: Reddish patches improved, but scales are still present in some areas.**
Treatment changes:	
- Azathioprine 2x50mg tablets by mouth daily.	
- Cetirizine 1x10mg tablets by mouth daily.	
- Desoximetasone cream 0.025% mixed with petroleum jelly (1:2) applied twice daily.	
- Pseudoceramide cream is applied frequently.	
Outcome	**After 2 weeks of azathioprine: Lesions improved with no side effects.**

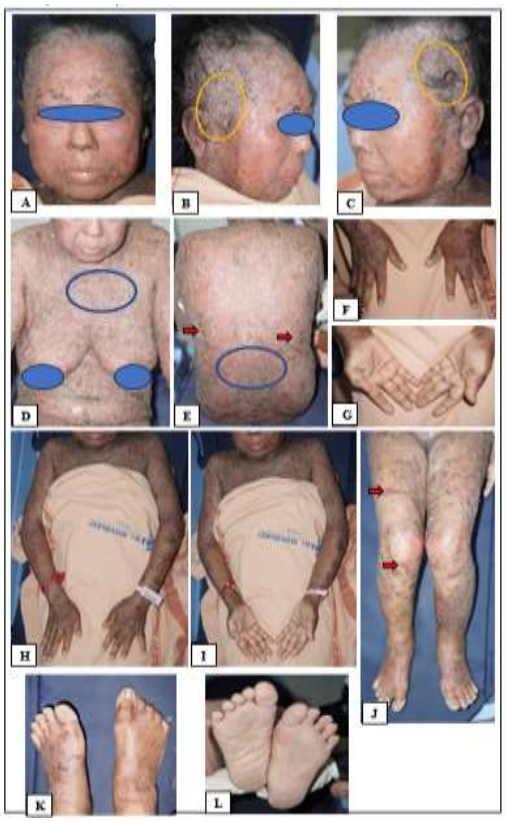

Figure 1. Dermatological status: In the generalized region, multiple erythematous patches are scattered discretely, some confluent, with scales on the surface (blue circle). Flaccid bullae that have ruptured leaving erosions in several areas are observed, with a positive Nikolsky sign (red arrow). The scalp region shows erythematous plaques with thick scales on the surface (yellow circle). No abnormalities are observed in the mucosa and nails.

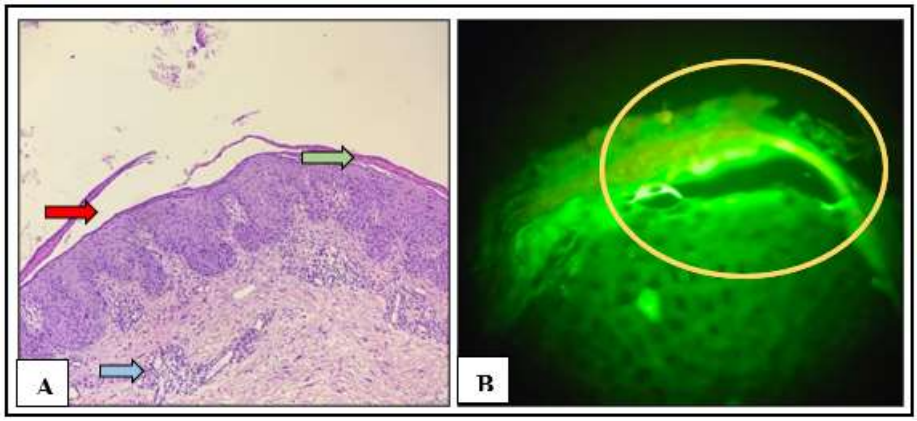

Figure 2. A. Histopathological examination revealed parakeratosis (green arrow), acantholysis, and a subcorneal cleft (red arrow) in the epidermis. Lymphocytic infiltration (blue arrow) was observed in the dermis with no evidence of malignancy (H&E, 100x). B. Direct immunofluorescence demonstrated positive IgG deposition (+) in the subcorneal layer (yellow circle).

3. Discussion

Pemphigus foliaceus (PF), an autoimmune blistering skin disease, hinges on the disruption of a critical protein known as desmoglein 1 (Dsg1). This disruption is orchestrated by autoantibodies, immune system components that mistakenly target the body's own tissues. To truly understand PF, we must delve deeper into the intricacies of Dsg1, its function, and how autoantibodies wreak havoc on skin integrity. Dsg1 is a transmembrane glycoprotein, a type of protein that spans the cell membrane and has sugar molecules attached. It belongs to the cadherin family, a group of proteins renowned for their role in cell-cell adhesion. In essence, cadherins act like molecular "velcro," sticking cells together to form tissues and organs. Within the epidermis, the outermost layer of the skin, Dsg1 plays a pivotal role in maintaining its structural integrity. Keratinocytes, the predominant cells in the epidermis, are bound together by desmosomes, specialized adhesive structures that act like "spot welds" between cells. Dsg1 is a key component of these desmosomes, ensuring that keratinocytes remain tightly connected, forming a cohesive and protective barrier against the external environment. In a healthy immune system, antibodies are produced to recognize and neutralize foreign invaders, such as bacteria and viruses. However, in autoimmune diseases like PF, the immune system goes awry, producing autoantibodies that mistakenly target the body's own tissues. In PF, these autoantibodies are specifically directed against Dsg1. The reasons for this misdirected immune response are not fully understood, but it is believed to involve a complex interplay of genetic predisposition and environmental triggers. The binding of autoantibodies to Dsg1 sets off a cascade of events that ultimately lead to the formation of blisters. Autoantibodies physically bind to Dsg1, blocking its ability to interact with other Dsg1 molecules on adjacent keratinocytes. This "steric hindrance" prevents the formation of proper cell-cell adhesions. The binding of autoantibodies can also trigger the internalization of Dsg1, meaning it is taken into the cell and degraded. This reduces the amount of Dsg1 available at the cell surface, further weakening cell-cell adhesion. Autoantibodies can interfere with intracellular signaling pathways that regulate desmosome assembly and function. This disruption can lead to the dismantling of desmosomes, compromising the structural integrity of the epidermis. The combined effects of steric hindrance, internalization, and signaling disruption lead to acantholysis, the loss of cell-cell adhesion between keratinocytes. As keratinocytes detach from each other, fluid

accumulates in the spaces between them, forming the characteristic superficial, fragile blisters seen in PF. The disruption of Dsg1 and the subsequent acantholysis manifest clinically as the hallmark features of PF. The blisters in PF are typically located in the upper layers of the epidermis, making them fragile and easily ruptured. When blisters rupture, they leave behind raw, eroded areas that often become covered with crusts. PF lesions can occur anywhere on the body, but they commonly affect the face, scalp, chest, and back. Unlike other forms of pemphigus, PF primarily affects the skin, with mucosal involvement being relatively rare. The severity and extent of PF can vary widely among individuals. Some may experience localized, mild outbreaks, while others may have widespread, severe disease that significantly impacts their quality of life. The lesions are often itchy and painful, and the chronic nature of the disease can take a toll on patients' physical and emotional well-being. Pemphigus foliaceus (PF) is a complex autoimmune disease, and despite extensive research, its exact cause remains elusive. However, a growing body of evidence points towards a multifactorial etiology, involving an intricate interplay between genetic predisposition and environmental triggers that ultimately converge to disrupt the delicate balance of the immune system. Genetic factors play a significant role in determining an individual's susceptibility to PF. Studies have consistently linked certain variations in human leukocyte antigen (HLA) genes to an increased risk of developing the disease. These genes, located on chromosome 6, are crucial for regulating the immune response. They encode proteins that help the immune system distinguish between "self" and "non-self," ensuring that the body's defenses are directed towards foreign invaders rather than its own tissues. In PF, specific HLA alleles, or gene variants, appear to increase the risk of developing autoimmunity against desmoglein 1 (Dsg1). These alleles may alter the way the immune system recognizes and processes Dsg1, leading to the production of autoantibodies that attack this essential cell adhesion protein. While HLA genes are strongly implicated in PF susceptibility, it's important to note that they are not the sole determinants. Other genes involved in immune regulation and skin barrier function are also likely to contribute to the overall genetic risk. While genetic predisposition lays the groundwork for PF, environmental triggers are often the "spark" that initiates the autoimmune cascade. These triggers can be diverse, ranging from medications and infections to ultraviolet radiation and even psychological stress. Certain medications have been reported to trigger PF in susceptible individuals. Penicillamine drug, used to treat rheumatoid arthritis and other conditions, has been associated with a relatively high risk of inducing PF. Captopril, An angiotensin-converting enzyme (ACE) inhibitor used to treat hypertension, captopril has also been linked to PF. Other medications, such as nonsteroidal anti-inflammatory drugs (NSAIDs) and certain antibiotics, have also been implicated in some cases. The exact mechanisms by which these medications trigger PF are not fully understood, but they may involve altering the structure of Dsg1, making it more susceptible to autoimmune attack, or disrupting immune regulation, leading to the production of autoantibodies. Infections, particularly viral infections, have also been suggested as potential triggers for PF. Viral infections can activate the immune system, potentially leading to a loss of self-tolerance and the production of autoantibodies. Exposure to ultraviolet (UV) radiation from sunlight or tanning beds may also contribute to the development of PF. UV radiation can cause DNA damage and alter immune responses in the skin, potentially triggering or exacerbating autoimmune reactions. Stress has been recognized as a potential trigger for various autoimmune diseases, including PF. Stress can disrupt the delicate balance of the immune system, potentially promoting autoimmunity. The precise mechanisms by which autoantibodies against Dsg1 disrupt cell-cell adhesion and lead to blister formation in PF are still being actively investigated. Autoantibodies binding to Dsg1 can physically obstruct the interaction between Dsg1 molecules on adjacent keratinocytes. This "steric hindrance"

prevents the formation of proper cell-cell adhesions, weakening the structural integrity of the epidermis. Autoantibody binding can trigger the internalization and subsequent degradation of Dsg1. This process reduces the amount of Dsg1 available at the cell surface, further compromising cell-cell adhesion. Autoantibodies can also interfere with intracellular signaling pathways that regulate desmosome assembly and function. Desmosomes are the specialized adhesive structures that anchor Dsg1 and other adhesion molecules to the cell cytoskeleton, providing mechanical strength to the epidermis. Disruption of these signaling pathways can lead to the dismantling of desmosomes, further contributing to acantholysis and blister formation. In some cases, autoantibodies may activate the complement system, a part of the innate immune system that can cause cell lysis and inflammation. Complement activation may contribute to the tissue damage seen in PF. The development of PF is likely a multi-step process, involving a combination of genetic susceptibility and environmental triggers that act in concert to disrupt immune tolerance and trigger autoimmunity against Dsg1. This concept is often referred to as the "multi-hit hypothesis." According to this hypothesis, individuals with a genetic predisposition to PF may require one or more environmental "hits" to trigger the disease. These hits can be cumulative, with each hit increasing the risk of developing PF. Once the threshold is crossed, the immune system loses its ability to tolerate Dsg1, leading to the production of autoantibodies and the characteristic clinical manifestations of PF. Pemphigus foliaceus (PF) presents a diagnostic challenge due to its variable clinical presentation and potential overlap with other skin disorders. However, a systematic approach combining clinical evaluation, histopathological examination, and immunofluorescence studies can lead to an accurate diagnosis and guide appropriate management strategies. A thorough clinical evaluation is the first step in diagnosing PF. The dermatologist will carefully examine the patient's skin, noting the distribution, appearance, and characteristics of the lesions. The presence of superficial, fragile blisters that rupture easily, leaving behind erosions and crusts, is suggestive of PF. The Nikolsky sign, where gentle rubbing of the skin causes separation of the epidermis, may also be positive in PF. A skin biopsy is essential for confirming the diagnosis of PF. The biopsy specimen is examined under a microscope to assess the microscopic features of the skin. In PF, the histopathological examination typically reveals acantholysis, the separation of keratinocytes within the epidermis, leading to the formation of intraepidermal blisters. Additionally, there may be inflammatory cell infiltrates, such as lymphocytes and eosinophils, within the epidermis and dermis. DIF is a specialized technique used to detect the presence and location of autoantibodies within the skin. In PF, DIF typically shows the deposition of IgG and sometimes C3 complement component in a characteristic intercellular pattern within the epidermis. This finding is highly specific for PF and helps distinguish it from other autoimmune blistering diseases. While not always necessary, IIF may be performed to detect circulating autoantibodies in the patient's serum. In PF, IIF typically shows the presence of autoantibodies that bind to the cell surface of keratinocytes. ELISA is another technique used to detect autoantibodies in the patient's serum. In PF, ELISA can specifically detect autoantibodies against desmoglein 1 (Dsg1). The management of PF often involves a combination of systemic and topical therapies aimed at suppressing the immune response, controlling inflammation, and promoting healing. Systemic corticosteroids, such as prednisone or methylprednisolone, are often the first-line treatment for PF. They work by suppressing the immune system and reducing inflammation. However, long-term use of corticosteroids can be associated with significant side effects, such as weight gain, osteoporosis, and increased risk of infections. In cases of severe or refractory PF, immunosuppressant medications, such as azathioprine, mycophenolate mofetil, or cyclophosphamide, may be added to the treatment regimen. These medications further suppress the immune system and can help reduce the

need for high-dose corticosteroids. However, they also carry potential risks, such as increased susceptibility to infections and other side effects. Other systemic therapies that may be considered in certain cases include intravenous immunoglobulin (IVIG), rituximab, and dapsone. Topical corticosteroids are often used to treat localized lesions in PF. They help reduce inflammation and itching. However, long-term use of potent topical corticosteroids can lead to skin thinning and other side effects. Other topical therapies that may be used include calcineurin inhibitors, such as tacrolimus or pimecrolimus, which have anti-inflammatory properties. Proper wound care is essential to prevent secondary infections and promote healing. This may involve gentle cleansing of the affected areas, application of topical antibiotics, and use of dressings to protect the skin. Pain management is important for improving the patient's quality of life. This may involve the use of over-the-counter pain relievers, such as acetaminophen or ibuprofen, or prescription pain medications in severe cases. PF can have a significant psychosocial impact due to the visible nature of the skin lesions. Patients may experience anxiety, depression, and social isolation. Providing emotional support and counseling can be beneficial. The prognosis of PF is generally favorable, with most patients responding well to treatment. However, PF can be a chronic condition that requires long-term management. The broken skin from blisters and erosions can become infected with bacteria or other pathogens. In some cases, PF can lead to scarring, particularly if the lesions are extensive or recurrent. The visible nature of the skin lesions can significantly impact a patient's self-esteem and quality of life. In severe cases with extensive skin involvement, patients may experience fluid and electrolyte imbalances due to loss of fluids through the skin. Regular follow-up with a dermatologist is essential to monitor the disease activity, adjust treatment as needed, and address any complications that may arise.[11-14]

Psoriasis vulgaris, often simply referred to as psoriasis, is a chronic inflammatory skin disease that affects millions of people worldwide. It is characterized by the presence of well-defined, erythematous plaques covered with silvery scales. These plaques can appear anywhere on the body, but they commonly occur on the elbows, knees, scalp, and lower back. While psoriasis is not contagious, its visible manifestations can significantly impact a person's quality of life, both physically and emotionally. Psoriasis is fundamentally an immune-mediated disease, meaning it arises from a dysfunction in the body's immune system. In healthy individuals, the immune system acts as a vigilant guardian, protecting the body from harmful invaders such as bacteria, viruses, and fungi. However, in psoriasis, the immune system mistakenly identifies components of the skin as foreign threats, triggering an inflammatory response that leads to the characteristic skin changes. At the cellular level, psoriasis involves a complex interplay of various immune cells, including T cells, dendritic cells, and macrophages. These cells release inflammatory mediators, such as cytokines and chemokines, that stimulate the proliferation of keratinocytes, the predominant cells in the epidermis. This accelerated keratinocyte turnover results in the thickening of the epidermis and the formation of raised, scaly plaques. Normally, keratinocytes mature and shed from the skin's surface in a controlled manner. However, in psoriasis, this process is significantly accelerated, leading to an accumulation of immature keratinocytes that form the characteristic silvery scales. Genetic factors play a significant role in determining an individual's susceptibility to psoriasis. Studies have identified numerous genes associated with an increased risk of developing the disease. These genes are involved in various aspects of immune regulation, skin barrier function, and inflammatory pathways. One of the most strongly associated genetic factors is the HLA-Cw6 allele, a variant of a gene involved in immune recognition. Other genes implicated in psoriasis include those encoding cytokines, such as IL-23 and IL-17, which play crucial roles in the inflammatory response. While genetic predisposition is a major contributor to psoriasis, it's important to

note that not everyone with a genetic predisposition will develop the disease. Environmental factors also play a crucial role in triggering or exacerbating psoriasis. A wide range of environmental factors can trigger or worsen psoriasis in genetically susceptible individuals. These triggers can vary from person to person, and identifying them is crucial for managing the disease effectively. Psychological stress is a well-known trigger for psoriasis flares. Stress can disrupt the delicate balance of the immune system, potentially promoting inflammation and exacerbating psoriasis symptoms. Infections, particularly streptococcal throat infections, have been linked to the onset and flares of psoriasis, especially guttate psoriasis, a form characterized by small, drop-shaped lesions. Injury to the skin, such as cuts, scrapes, or sunburn, can trigger the Koebner phenomenon, where new psoriasis lesions develop at the site of injury. Certain medications, including lithium, beta-blockers, and antimalarial drugs, have been associated with triggering or worsening psoriasis. Smoking is a significant risk factor for psoriasis and can also make the disease more difficult to treat. Excessive alcohol consumption can also exacerbate psoriasis symptoms. Cold, dry weather can worsen psoriasis symptoms, while warm, sunny weather may improve them. The most common form, characterized by well-defined, erythematous plaques covered with silvery scales. Characterized by small, drop-shaped lesions that often appear suddenly, commonly following a streptococcal infection. Occurs in skin folds, such as the armpits, groin, and under the breasts, and appears as smooth, red patches. Characterized by pus-filled blisters, often accompanied by fever and malaise. A severe form involving widespread redness and scaling of the skin. In addition to skin lesions, psoriasis can also affect the nails, causing pitting, discoloration, and thickening. Psoriatic arthritis, a form of arthritis that can accompany psoriasis, affects the joints, causing pain, stiffness, and swelling. Psoriasis can significantly impact a person's quality of life. The visible nature of the skin lesions can lead to self-consciousness, embarrassment, and social isolation. The itching and pain associated with psoriasis can also disrupt sleep and daily activities. Furthermore, psoriasis is associated with an increased risk of other health conditions, such as cardiovascular disease, diabetes, and depression.[15-17]

Pemphigus foliaceus (PF) and psoriasis vulgaris are both chronic inflammatory skin diseases with immune system involvement at their core. However, they are distinct entities with different underlying mechanisms. PF is characterized by autoantibodies targeting desmoglein 1 (Dsg1), leading to acantholysis and the formation of superficial blisters. Psoriasis, on the other hand, is driven by T-cell mediated inflammation and results in hyperproliferation of keratinocytes, leading to the formation of scaly plaques. Despite these distinct pathophysiological pathways, an intriguing association between PF and psoriasis vulgaris has been observed. Several case reports and studies have documented the development of PF in patients with a history of psoriasis, raising questions about the nature of this relationship and the potential underlying mechanisms. The association between PF and psoriasis vulgaris is undoubtedly rare, but its occurrence raises the possibility of a causal link. Could psoriasis predispose individuals to develop PF? Or could shared genetic or environmental factors contribute to the development of both conditions? Both PF and psoriasis vulgaris have a strong genetic component. Specific HLA genes have been associated with an increased risk of both conditions. It is plausible that shared genetic susceptibility loci may contribute to the co-occurrence of these diseases. For instance, certain HLA alleles may influence immune regulation in a way that increases the risk of both T-cell mediated inflammation in psoriasis and autoantibody production against Dsg1 in PF. Psoriasis vulgaris is characterized by a dysregulated immune response, with an imbalance between pro-inflammatory and anti-inflammatory cytokines. This altered immune milieu may create conditions favorable for the development of autoimmunity, including the production of autoantibodies against

Dsg1. The chronic inflammation in psoriasis could potentially "prime" the immune system, making it more prone to lose tolerance to self-antigens and initiate an autoimmune attack against Dsg1. Certain environmental triggers, such as medications, infections, and ultraviolet radiation, have been implicated in both PF and psoriasis vulgaris. Exposure to these triggers may exacerbate both conditions or even trigger the development of one condition in individuals with a predisposition to the other. For example, certain medications, such as lithium and beta-blockers, have been associated with both psoriasis and PF. Similarly, infections, particularly viral infections, may trigger or exacerbate both conditions. Both PF and psoriasis vulgaris involve disruption of the epidermal barrier, the outermost layer of the skin that protects the body from external insults. This barrier dysfunction may contribute to the development of both conditions or facilitate the entry of environmental triggers that can exacerbate inflammation. A compromised epidermal barrier may allow for the entry of antigens that can trigger an immune response, potentially leading to the development of autoimmunity in susceptible individuals. The exact mechanisms underlying the association between PF and psoriasis vulgaris remain unclear. However, a growing body of research is providing insights into the complex interplay of genetic, immunological, and environmental factors that may contribute to this intriguing connection. Studies have shown that T cells, a type of immune cell that plays a central role in psoriasis, may also be involved in the pathogenesis of PF. In psoriasis, T cells infiltrate the skin and release inflammatory cytokines that drive keratinocyte hyperproliferation. In PF, T cells may contribute to the loss of tolerance to Dsg1 and the production of autoantibodies. An imbalance in cytokine levels, particularly an increase in pro-inflammatory cytokines such as IL-17 and TNF-alpha, has been observed in both PF and psoriasis vulgaris. These cytokines may contribute to the inflammation and tissue damage seen in both conditions. Genetic studies have identified polymorphisms, or variations, in genes involved in immune regulation and skin barrier function that may increase the risk of both PF and psoriasis vulgaris. These polymorphisms may affect the expression or function of proteins involved in immune responses and skin barrier integrity. Epigenetic modifications, which are changes in gene expression that do not involve alterations in the DNA sequence, may also play a role in the association between PF and psoriasis vulgaris. These modifications can be influenced by environmental factors and may contribute to the development of both conditions. The association between PF and psoriasis vulgaris has important clinical implications. Dermatologists should be aware of this potential overlap and consider the possibility of PF in patients with psoriasis who present with new blistering lesions. Early diagnosis and appropriate treatment are crucial for managing PF and preventing complications. In some cases, the clinical presentation of PF in patients with psoriasis may be atypical, making diagnosis more challenging. The presence of both scaly plaques and blisters may create a confusing clinical picture. A thorough history, physical examination, and skin biopsy are essential for accurate diagnosis.[18-20]

4. Conclusion

This case report presents a rare instance of PF arising in a patient with a history of psoriasis vulgaris. The development of PF in this patient highlights the potential for complex interactions between these two conditions, even though they have distinct pathophysiological mechanisms. The patient's clinical presentation, with the development of fragile blisters and erosions, along with the histopathological findings of acantholysis and the positive IgG deposition on direct immunofluorescence, confirmed the diagnosis of PF. The patient's treatment with systemic corticosteroids and azathioprine led to a significant improvement in her skin lesions, underscoring the importance of appropriate treatment in managing PF and improving patient outcomes. This case underscores the need for increased awareness among healthcare professionals about the potential

association between PF and psoriasis vulgaris. Further research is needed to fully elucidate the underlying mechanisms and identify potential risk factors for this rare but intriguing clinical scenario.

5. References

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