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COMBINATION THERAPY WITH CORTICOSTEROIDS AND MYCOPHENOLATE MOFETIL IN THE MANAGEMENT OF PEMPHIGUS VULGARIS: A CASE REPORT

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ABSTRACT
Pemphigus vulgaris (PV) is a rare, potentially life-threatening autoimmune blistering disease caused by IgG autoantibodies against desmoglein proteins, leading to acantholysis and intraepidermal blister formation. The incidence of PV ranges from 0.76 new cases per million population per year in Finland to approximately 3.5 new cases per million population per year in other regions. Early diagnosis and appropriate immunosuppressive therapy are crucial to prevent disease progression and complications. This study aims to report the clinical presentation, diagnostic approach, and therapeutic outcome of combination therapy using systemic corticosteroids and mycophenolate mofetil in a patient with pemphigus vulgaris. This is a descriptive case report of a 40-year-old female diagnosed with pemphigus vulgaris. Clinical evaluation, histopathological examination, and therapeutic response were documented and analyzed. A 40-year-old woman who presented with a one-week history of painful, generalized flaccid bullae and erosions involving the face, scalp, trunk, and extremities. Histopathological examination revealed suprabasal intraepidermal blistering with a characteristic row of tombstones appearance, confirming the diagnosis of pemphigus vulgaris.Initial treatment with systemic corticosteroids resulted in partial clinical improvement; however, new lesions continued to develop. The addition of mycophenolate mofetil as a steroid-sparing agent led to significant disease control, with cessation of new blister formation and gradual healing of existing lesions. The patient showed marked clinical improvement with good functional and vital prognosis, although long-term remission remains uncertain. This case highlights a severe and extensive presentation of pemphigus vulgaris and emphasizes the importance of clinicopathological correlation for accurate diagnosis. Early combination therapy with systemic corticosteroids and mycophenolate mofetil may be effective in controlling refractory disease and minimizing corticosteroid-related adverse effects.

Keywords: autoimmune blistering disease; mycophenolate mofetil; pemphigus vulgaris; systemic corticosteroids

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INTRODUCTION

Pemphigus vulgaris (PV) is a rare but potentially life-threatening chronic autoimmune blistering disease characterized by the formation of intraepidermal vesicles and bullae affecting the skin and mucous membranes. The disease is caused by immunoglobulin G (IgG) autoantibodies targeting desmosomal proteins, particularly desmoglein 1 and 3, leading to acantholysis and loss of intercellular adhesion between keratinocytes. Clinically, PV often begins with painful oral mucosal lesions before progressing to involve the skin, significantly impairing patients' quality of life (Baroukhian et al., 2025).

The incidence of PV varies geographically, ranging from less than 1 case per million population per year in some regions to higher rates in specific populations. Recent epidemiological studies report a global pooled incidence of approximately 2.83 cases per million person-years, with higher rates

observed in regions such as South Asia and the Middle East (Zhao et al., 2023; Stone et al., 2024). PV remains the most common subtype of pemphigus, with increased prevalence among individuals of Mediterranean and Middle Eastern descent (Welc et al., 2024). Although PV can occur at any age, disease onset most commonly occurs between 40 and 60 years (Ingold, 2024; Rosi-Schumacher et al., 2023).

Pathophysiologically, PV is driven by IgG autoantibodies directed primarily against desmoglein 1 (Dsg1) and desmoglein 3 (Dsg3), key components of desmosomes on keratinocyte surfaces. Disruption of these proteins leads to loss of cell adhesion and subsequent blister formation. Patients with anti-Dsg3 autoantibodies typically present with mucosal lesions, whereas those with both anti-Dsg1 and anti-Dsg3 antibodies develop mucocutaneous involvement. Diagnosis is established through Tzanck smear, histopathological examination, and direct immunofluorescence, which remains the gold standard (Murrell et al., 2020).

Despite its rarity, PV management remains challenging due to its chronic course, risk of complications, and long-term treatment-related adverse effects. Systemic corticosteroids remain the mainstay of therapy due to their rapid immunosuppressive effects (Zhang et al., 2026). Oral prednisolone is typically administered at a dose of 0.5–1.0 mg/kg/day, with dose escalation in refractory cases until disease control is achieved, followed by gradual tapering after the consolidation phase (Tran & Walter, 2023). However, prolonged use of high-dose corticosteroids is associated with significant adverse effects, including diabetes mellitus, hypertension, osteoporosis, and increased risk of infection, necessitating additional therapeutic strategies to reduce steroid dependence (Earlia et al., 2026).

In recent years, immunosuppressive agents have increasingly been used as adjuvant therapy to achieve a steroid-sparing effect. One commonly used agent is mycophenolate mofetil (MMF), which inhibits lymphocyte proliferation by blocking inosine monophosphate dehydrogenase. Combination therapy with corticosteroids and MMF has been reported to effectively control disease activity, accelerate remission, and reduce steroid-related adverse effects (Zhang et al., 2026; Almousa et al., 2025). Several case reports and observational studies have demonstrated that this combination provides significant clinical improvement, particularly in patients with comorbidities or limited therapeutic options (Earlia et al., 2026). Moreover, recent trends in PV management suggest that combination therapy is preferred over monotherapy, especially in moderate to severe cases, to achieve optimal disease control (Bagchi et al., 2026).

Therefore, this study aims to describe the clinical outcomes and therapeutic effectiveness of combination therapy with corticosteroids and mycophenolate mofetil in the management of pemphigus vulgaris, and to provide insight into its role as a steroid-sparing strategy in clinical practice.

METHOD

This study is presented as a single case report. The data were obtained through direct clinical observation, patient anamnesis, physical examination, laboratory investigations, and histopathological evaluation. The patient was selected purposively as she presented with new-onset erythroderma following Sambiloto consumption and no other identifiable triggers. Data collection included clinical photographs, laboratory results, and skin biopsy findings. The information was descriptively analyzed by comparing the patient's presentation with existing literature on drug-induced erythroderma and adverse reactions to herbal medicines.

RESULT

A 40-year-old Batak woman presented to the Emergency Department of H. Adam Malik General Hospital with a chief complaint of painful fluid-filled blisters and widespread skin peeling that had developed over the past week. Initially, the blisters appeared on her back and progressively increased in number, spreading to the face, neck, chest, and scalp. The blisters were flaccid and easily ruptured, releasing clear fluid and leaving erythematous patches.Prior to the onset of the skin lesions, the patient had received treatment at a primary healthcare center for fever; however, she was unable to recall the medications prescribed. She had also sought care from a general practitioner but did not remember the medications taken, and her symptoms did not improve. She denied the use of other medications. There was no history of topical application before or after the onset of the lesions.

Regarding her past medical history, the patient had never experienced a similar condition. She denied any history of systemic diseases such as diabetes mellitus, hypertension, or asthma. There was no history of food or drug allergies. A family history of similar illness was also denied. On physical examination, the patient appeared weak but was fully conscious (compos mentis). Her vital signs were as follows: blood pressure 100/70 mmHg, respiratory rate 20 breaths per minute, pulse rate 90 beats per minute, axillary temperature 37.2°C, Visual Analog Scale (VAS) pain score of 6, and body weight 70 kg.

General examination revealed a normocephalic head. Ocular examination showed no signs of anemia or icterus. Examination of the ears, nose, and throat was unremarkable, and there was no cervical lymphadenopathy. Dermatological examination revealed flaccid bullae on an erythematous base, along with erythematous macules, erosions, excoriations, and crusts distributed over the facial region, right side of the neck, thoracic region, and scalp (Figure 1).

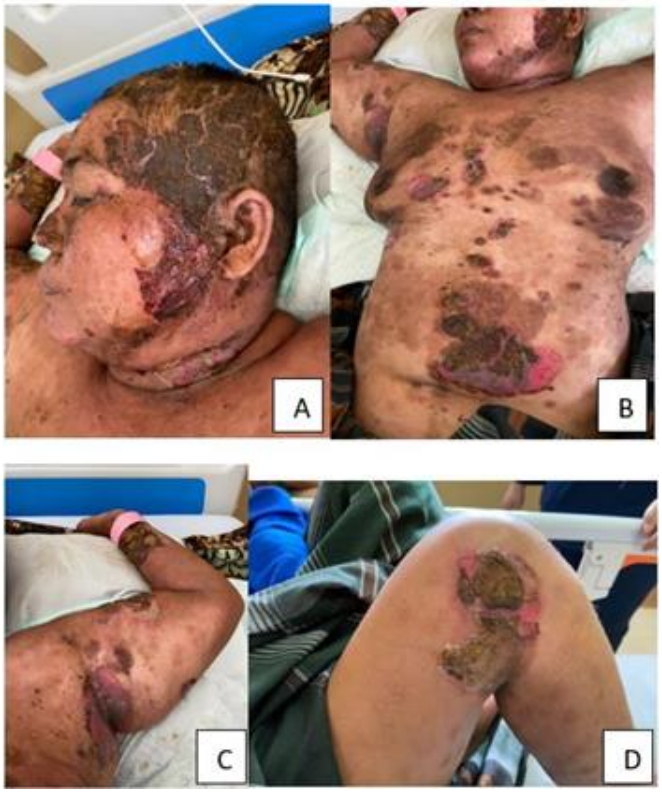

Figure 1. Flaccid bullae on an erythematous base, accompanied by erosions, excoriations, and crusts involving the scalp, facial region, neck, anterior thoracic region, abdomen, and bilateral brachial regions (a–d).

Routine laboratory blood examination revealed leukocytosis (white blood cell count: 12.1 × 10^3/µL), while other hematological parameters were within normal limits. Random blood glucose

levels were within normal limits. Liver and renal function tests were also within normal limits. Histopathological examination showed the following findings: On gross examination, a single skin tissue specimen approximately the size of a mung bean was received. The specimen was firm in consistency and grayish-black in color. Microscopically, the tissue section was derived from the skin and showed stratified squamous epithelium composed of cells with round to oval nuclei, fine chromatin, and eosinophilic cytoplasm. Hyperkeratosis and acantholysis were observed. An intraepidermal blister located in the suprabasal layer was identified, forming the characteristic "row of tombstones" appearance. In the subepidermal layer, the stroma consisted of fibrocollagenous connective tissue with lymphocytic inflammatory cell infiltration (Figure 2). No evidence of malignancy was identified in the specimen. The histopathological findings were consistent with pemphigus vulgaris.

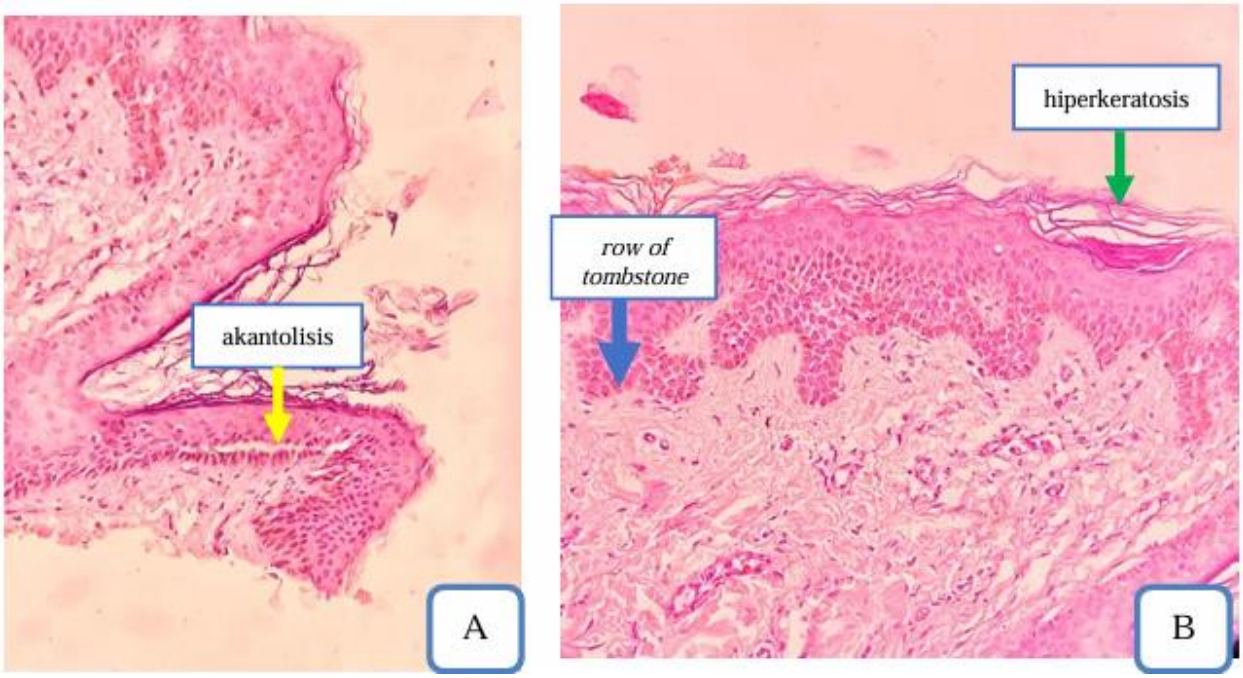

Figure 2. Histopathological findings. Stratified squamous epithelium composed of cells with round to oval nuclei, fine chromatin, and eosinophilic cytoplasm showing hyperkeratosis and acantholysis (A). An intraepidermal blister located in the suprabasal layer forming the characteristic "row of tombstones" appearance is also observed (B).

The patient was admitted for inpatient care and co-managed with the Department of Internal Medicine. Treatment initiated by the Internal Medicine team included intravenous fluid therapy with 0.9% sodium chloride at a rate of 20 drops per minute and ceftriaxone 1 g administered intravenously every 12 hours. A consultation was also requested from the Department of Dermatology and Venereology. Based on the recommendations from the Dermatology and Venereology team, additional therapy was initiated, consisting of intravenous methylprednisolone 31.25 mg every 12 hours and intravenous ranitidine 50 mg every 12 hours. Wet compresses with 0.9% sodium chloride solution were applied for 15–20 minutes every 4 hours to the moist wounds (erosions) and bullae on the facial, cervical, and thoracic regions. Fusidic acid (Fuson®) cream was applied twice daily to the affected skin areas.

At the 7-day follow-up, the skin lesions were still desquamating and partially covered with crusts; however, the amount of crusting had decreased, and no new bullae were observed. Intravenous methylprednisolone at a dose of 31.25 mg every 12 hours was continued for the following 7 days. Subsequently, therapy was transitioned to oral methylprednisolone at an equivalent dose, followed by gradual tapering. Topical therapy was maintained. (Figure 3).

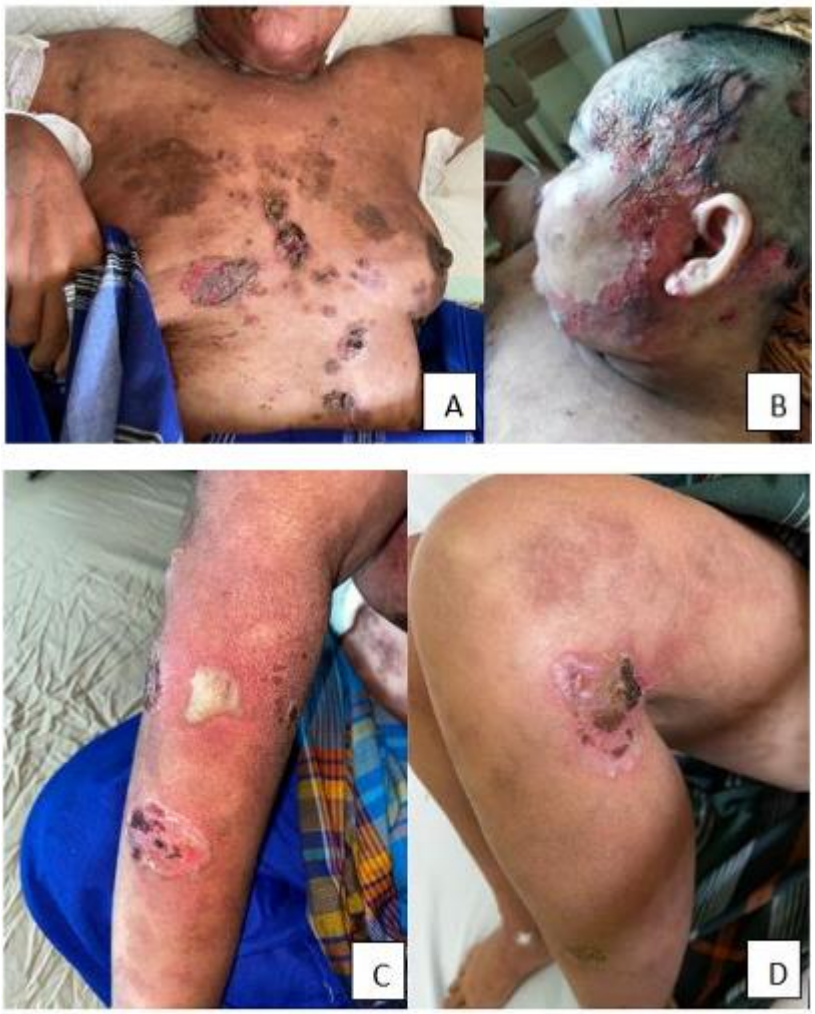

Figure 3. First follow-up. Multiple flaccid bullae over generalized regions had slightly decreased in number; however, flaccid bullae were still observed on the upper extremities (a–d).

At the second follow-up, 14 days after initiation of treatment, recurrent blisters were observed on the face. These lesions had ruptured and formed crusts. New fluid-filled bullae appeared on the right thigh; several had ruptured, while some lesions across the body had dried. The patient also complained of generalized weakness and recurrent epigastric discomfort. Pain persisted at the time of evaluation (Figure 4). The management provided at this stage included mycophenolate mofetil 1 g twice daily, methylprednisolone 16 mg three times daily, and omeprazole 20 mg twice daily. Wet compresses with 0.9% sodium chloride solution were applied for 15 minutes six times daily. Fusidic acid cream was applied twice daily to the affected areas.

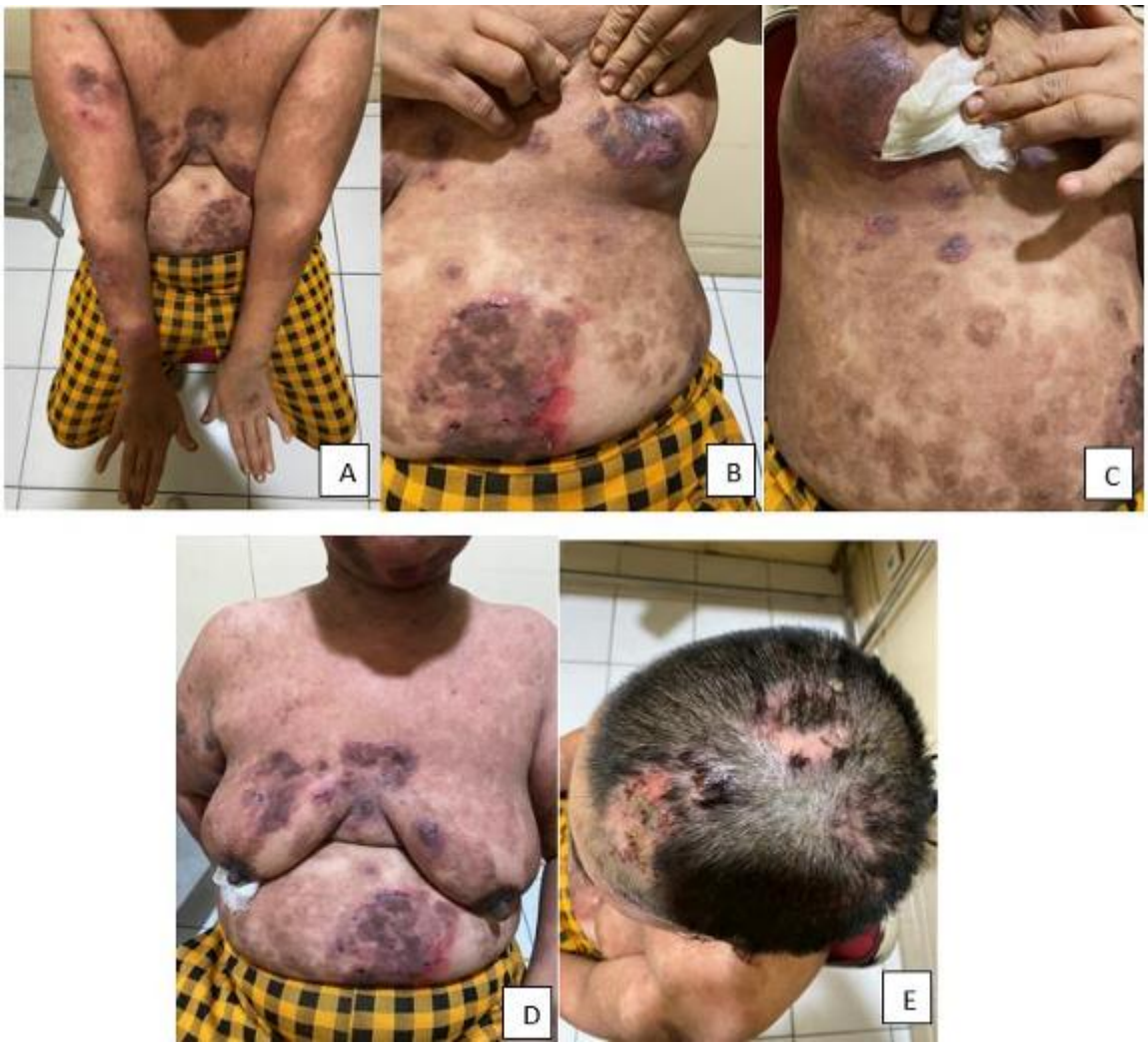

Figure 4. Second follow-up. Erosions and excoriations were observed in the facial region. Multiple flaccid bullae over generalized areas had slightly decreased in number; however, new flaccid bullae were still present on the chest and upper extremities (a–e).

At the third follow-up visit, the patient reported clinical improvement. Facial lesions had markedly decreased. The fluid-filled blisters on the arms had dried, and residual lesions on the body were resolving with post-inflammatory hyperpigmentation. The patient no longer complained of pain (Figure 5).The treatment regimen was continued, consisting of mycophenolate mofetil 1 g twice daily, methylprednisolone 16 mg three times daily, and omeprazole 20 mg twice daily. Wet compresses with 0.9% sodium chloride solution were applied for 15 minutes six times daily, and fusidic acid cream was applied twice daily to the affected areas. Based on the history, physical examination, and supporting investigations, the prognosis in this patient was considered *quo ad vitam ad bonam* (regarding life expectancy), *quo ad functionam ad bonam* (regarding functional outcome), *quo ad sanationam dubia*. (regarding complete recovery).

Figure 5. Third follow-up. Erosions and excoriations in the facial region had decreased (a). Multiple flaccid bullae over generalized areas had further reduced in number compared to previous visits (a–d).

DISCUSSION

Cutaneous manifestations of pemphigus vulgaris (PV) typically present as flaccid vesicles or bullae on an erythematous base. These lesions may involve the entire body and are often accompanied by pain. The bullae are fragile and rupture easily, resulting in erosions followed by crust formation that may persist for an extended period. Healing generally occurs with post-inflammatory hyperpigmentation and without scar formation. Bullae may arise on either normal-appearing skin or erythematous skin (Ingold, 2024; Kridin, 2018).

Histopathologically, pemphigus vulgaris (PV) is characterized by a suprabasal intraepidermal blister. The initial pathological change involves intercellular edema, followed by detachment of suprabasal keratinocytes from the basal cells, leading to cleft formation and blister development. The basal cells remain attached to the basement membrane but lose cohesion with adjacent keratinocytes, resulting in the characteristic “row of tombstones” appearance at the base of the blister (Ingold, 2024)

Bullous pemphigoid was excluded as a differential diagnosis because its lesions typically present as tense vesicles or bullae filled with clear or occasionally hemorrhagic fluid. Histologically, bullous pemphigoid is characterized by subepidermal blister formation accompanied by a prominent inflammatory infiltrate rich in eosinophils (Obijiofor et al., 2024; Saal et al., 2025). Stevens–Johnson syndrome (SJS) was also ruled out, as it is typically preceded by prodromal symptoms such as fever and malaise and initially presents with macules that progress to papules, vesicles, or bullae,

which subsequently rupture. The lesions commonly involve the palms, soles, extensor surfaces, and mucous membranes. Histopathological findings usually demonstrate epidermal necrosis with minimal inflammatory cell infiltration, along with changes at the dermoepidermal junction ranging from vacuolar alteration to subepidermal blister formation (Harr & French, 2010)

The primary goal of therapy in the management of pemphigus vulgaris is to prevent the formation of new lesions and to promote healing of existing lesions. Treatment is considered successful when it effectively suppresses the production of pathogenic autoantibodies. The choice of therapy depends on the severity of the disease. Additional factors influencing therapeutic decisions include patient-related considerations and drug-related factors. In the absence of absolute contraindications, systemic glucocorticoids remain the first-line initial therapy (Amagai, 2018).

The initial recommended dose of prednisone in pemphigus vulgaris is 0.5–1.0 mg/kg/day, depending on disease severity. The dose may be adjusted based on clinical response, and gradual tapering is initiated once disease control is achieved to minimize corticosteroid-related adverse effects (Schmidt et al., 2019). After two weeks of treatment, the patient showed no significant improvement, and new bullae continued to appear on the face, palms, and thighs. Therefore, additional therapy with mycophenolate mofetil 1 g twice daily was initiated as a steroid-sparing agent. After one week of combination therapy, no new bullae developed, and clinical improvement was observed.

Long-term administration of systemic corticosteroids is associated with significant adverse effects, including diabetes mellitus, hypertension, osteoporosis, and metabolic disturbances. Therefore, combination therapy with immunosuppressive agents is recommended to reduce cumulative corticosteroid exposure and minimize treatment-related toxicity (Wang et al., 2024; Earlia et al., 2025).

The rapid therapeutic effect of corticosteroids is attributed to their potent anti-inflammatory and immunosuppressive actions, which help counteract autoantibody-mediated disruption of keratinocyte adhesion. However, corticosteroid-related adverse events may contribute to increased morbidity and mortality, highlighting the importance of optimizing treatment strategies and identifying the minimum effective dose required to induce and maintain remission (Christjaroon et al., 2025; Zeng et al., 2022).

Azathioprine and mycophenolate mofetil are widely used as first-line adjuvant immunosuppressive therapies in pemphigus and are considered effective steroid-sparing agents. Azathioprine has demonstrated efficacy in reducing corticosteroid requirements when used in combination therapy, while mycophenolate mofetil is associated with favorable tolerability and effective disease control, making it a commonly preferred alternative in clinical practice (Schmidt et al., 2019)

The recommended dose of azathioprine in pemphigus vulgaris is approximately 1–3 mg/kg/day administered orally, with potential adverse effects including myelosuppression and hepatotoxicity. In contrast, mycophenolate mofetil is generally associated with fewer hepatic adverse effects and a more favorable safety profile (Schmidt et al., 2019). The severity and clinical course of pemphigus vulgaris vary considerably. Mortality is highest in the early years following disease onset; however, prognosis improves significantly with early diagnosis and appropriate treatment. Poor prognostic factors include advanced age, extensive disease involvement, and the presence of comorbidities (Schmidt et al., 2021; Rosi-Schumacher et al., 2023).

CONCLUSION

This case underscores a rare and severe presentation of pemphigus vulgaris with extensive generalized involvement, emphasizing the critical role of histopathological confirmation in

establishing an accurate diagnosis. The addition of mycophenolate mofetil as a steroid-sparing agent resulted in rapid disease control after an inadequate response to systemic corticosteroids alone.

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