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From Blister to Breakthrough: A Case Report on Pemphigus Vulgaris with Early Rituximab Intervention

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ABSTRACT

Background:
Pemphigus vulgaris (PV) is a rare, chronic autoimmune blistering disorder characterized by IgG autoantibodies targeting desmoglein-3. This results in loss of keratinocyte adhesion and intraepidermal blistering. While systemic corticosteroids have historically formed the cornerstone of therapy, long-term use is associated with significant morbidity. Rituximab, an anti-CD20 monoclonal antibody has recently emerged as a first-line agent due to its superior efficacy and safety profile. Early intervention with targeted immunotherapy is critical in improving disease outcomes and minimizing complications.

Case Report:
We report the case of a 50-year-old woman who presented with a one-month history of painful flaccid bullae and erosive, crusted lesions predominantly affecting the trunk, with extension to the chest, neck, scalp, ears, and oral cavity. Histopathological analysis revealed suprabasal clefting with acantholysis, and direct immunofluorescence demonstrated intercellular IgG deposition in a fishnet pattern, confirming the diagnosis of PV. The patient was started on rituximab as primary systemic therapy, combined with oral prednisolone, topical clobetasol, and dexamethasone elixir as adjunctive treatment. Supportive wound care and prophylactic supplementation with calcium and vitamin D were also provided. At two-week follow-up, the patient demonstrated reduced blister formation, improved healing of erosions, and no treatment-related complications.

Conclusion:
This case highlights the importance of early diagnosis and initiation of rituximab-based therapy in pemphigus vulgaris. Early intervention with B-cell–directed immunotherapy, combined with corticosteroids and supportive care, can reduce disease activity, limit steroid exposure, and improve long-term prognosis. A multidisciplinary, patient-centred approach remains essential to optimize outcomes in this potentially life-threatening disease.

Keywords: Autoimmune blistering diseases, Corticosteroids, Pemphigus vulgaris, Rituximab, Skin biopsy

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INTRODUCTION

Pemphigus vulgaris (PV) is a chronic, potentially life-threatening autoimmune blistering disorder characterized by the production of pathogenic IgG autoantibodies directed against desmoglein-3, a critical component of desmosomes.[1] These intercellular junctions mediate adhesion between keratinocytes in the epidermis and mucosal epithelium. Autoantibody-mediated disruption of desmosomes results in acantholysis-loss of cohesion between adjacent keratinocytes-leading to the formation of fragile, flaccid intraepidermal blisters affecting both the skin and mucous membrane.[2]

Although PV can occur at any age, it most commonly affects individuals in middle adulthood, with no significant sex predilection. A recent systematic review and meta-analysis estimated the global incidence of PV at approximately 2.83 cases per million person-years (95% CI, 2.14-3.61).[3] Early diagnosis is essential, as timely therapeutic intervention can significantly alter disease progression and improve clinical outcomes.[4]

The primary therapeutic objective in PV is to induce and maintain remission, defined by complete re-epithelialization of lesions, absence of new blister formation, and the ability to taper

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immunosuppressive therapy without relapse. Advances in understanding PV pathogenesis have underscored the role of B lymphocytes in sustaining autoimmunity. As such, targeted B-cell therapies have emerged as promising strategies for disease control.[5]

Historically, systemic corticosteroids have formed the cornerstone of PV treatment. While effective, their long-term use is associated with significant adverse effects, including opportunistic infections, metabolic disturbances, and end-organ damage. The introduction of rituximab, a monoclonal antibody targeting the CD20 antigen on B cells, has transformed the management of PV. Rituximab has demonstrated superior efficacy compared to mycophenolate mofetil (MMF) in randomized controlled trials and is now recommended as a first-line agent due to its sustained clinical responses and favorable safety profile.[6]

This report presents the case of a 50-year-old woman diagnosed with PV, detailing her clinical presentation, diagnostic evaluation, and therapeutic course. It highlights the importance of early recognition and a multidisciplinary approach in optimizing patient outcomes and minimizing disease-related complications.

CASE REPORT

A 50-year-old woman with no significant past medical history presented to the dermatology clinic with a one-month history of painful, flaccid blisters and crusted lesions primarily affecting the trunk. The lesions initially appeared as small vesicles that progressively enlarged, ruptured, and formed painful erosions with overlying crust. The patient reported marked discomfort, particularly during movement, with the most prominent involvement noted on the upper and lower abdominal regions.

On clinical examination, multiple flaccid bullae and widespread areas of crusted erosions were observed across the trunk. Additional lesions were noted on the chest, abdomen, neck, scalp, ears, and oral cavity. No active mucosal lesions were evident at the time of examination. The remainder of the cutaneous and systemic examination was unremarkable. The patient was referred to the Family Medicine Department for further laboratory evaluation and skin biopsy.

Baseline laboratory investigations, including complete blood count, renal and hepatic function panels, and serum electrolytes, were within normal reference ranges, effectively excluding systemic or infectious etiologies of blistering.

Two punch biopsies were obtained from the perilesional skin of active bullous lesions on the upper and lower abdomen. The first specimen, measuring 0.5 cm in diameter and 0.4 cm in depth, was submitted in formalin for routine histopathological analysis. The second was preserved in Michel's transport medium for direct immunofluorescence (DIF) studies.

Histopathological examination revealed an intraepidermal blister with suprabasal clefting and prominent acantholysis. Clusters of acantholytic keratinocytes were noted within the blister cavity, and the adjacent epidermis demonstrated keratinocyte dissociation. The underlying dermis showed a mild to moderate superficial perivascular lymphocytic infiltrate (Fig 1).

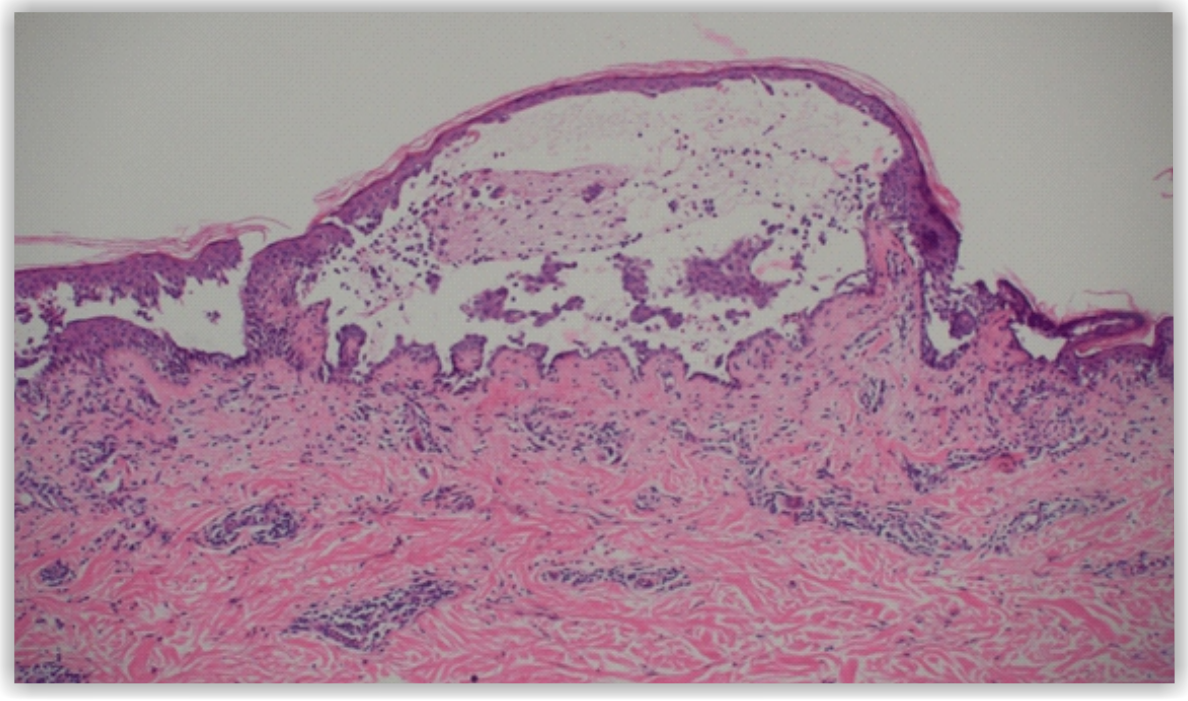

Figure 1: Histopathological features of pemphigus vulgaris in a punch biopsy stained with hematoxylin and eosin (H&E).

Direct immunofluorescence (DIF) studies were performed on perilesional skin biopsies to confirm the diagnosis. Intercellular deposition of IgG in a characteristic fishnet pattern with focal disruption and early acantholysis, indicating loss of keratinocyte adhesion in affected areas were seen (Fig.2). In contrast more uniform intercellular IgG staining with preserved epidermal architecture, consistent with early-stage pemphigus vulgaris (Fig.3). These findings supported the presence of autoantibodies targeting desmosomal proteins, primarily desmoglein 3, and confirmed the diagnosis of pemphigus vulgaris. The variation in staining patterns between the two samples reflects the dynamic nature and progression of the disease across different biopsy sites.

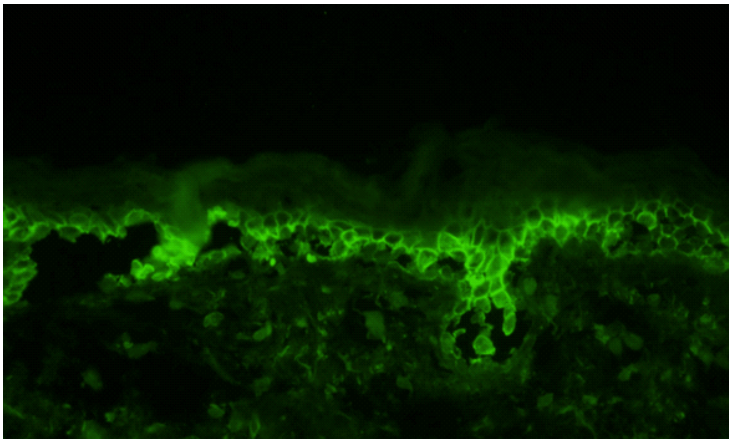

Figure 2: Direct immunofluorescence showing intercellular IgG with early acantholysis in pemphigus vulgaris

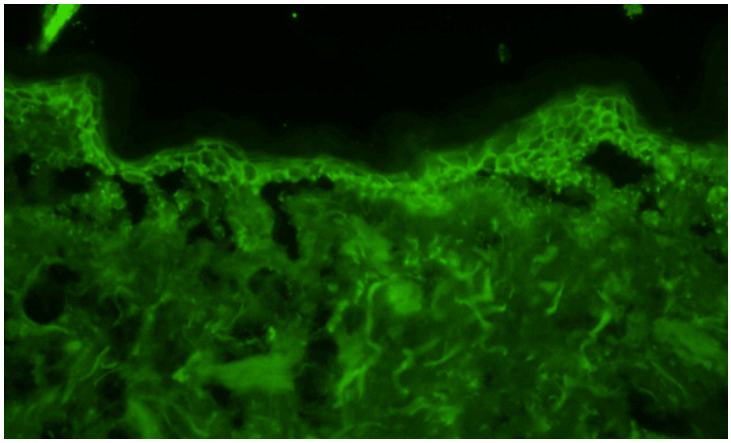

Figure 3: Direct immunofluorescence showing classic fishnet IgG staining in pemphigus vulgaris

Following diagnosis, the patient was initiated on a treatment regimen aimed at immunosuppressive control and rapid symptom resolution. Rituximab was administered as the primary systemic therapy to target CD20- positive B lymphocytes, thereby reducing autoantibody production. This approach was selected to reduce the need for prolonged corticosteroid exposure.

Systemic corticosteroids were introduced with prednisolone 50 mg orally once daily to control acute inflammation. A tapering schedule was planned as rituximab's effects became clinically evident. Dexamethasone elixir (0.5 mg/5 mL) was used during the early treatment phase to bridge the time until rituximab's full therapeutic onset and to facilitate tapering of systemic steroids.

Topical clobetasol propionate 0.05% cream was prescribed for twice-daily application to affected skin areas, offering localized anti-inflammatory effects and promoting re-epithelialization. The patient received detailed wound care instructions to minimize the risk of secondary infection and enhance healing. She was counseled about the potential adverse effects of corticosteroids, including the risk of osteoporosis. As a preventive measure, calcium and vitamin D supplementation was initiated.

A follow-up visit was scheduled two weeks after the start of therapy. At review, the patient reported a reduction in pain and a decrease in new blister formation. While some lesions persisted, their severity had lessened. Repeat blood tests remained within normal parameters, and no treatment-related

DISCUSSION

Pemphigus vulgaris is an autoimmune blistering disorder that predominantly affects the skin and mucous membranes. Left untreated, it carries significant morbidity and mortality . PV is characterized by the formation of thin-walled, fragile blisters that rupture easily, leaving painful, denuded areas. A key clinical finding is a positive Nikolsky's sign, where gentle lateral pressure on the skin causes epidermal detachment or blister extension.[7]

Histopathological analysis remains critical for diagnosis. The pathogenesis of PV involves autoantibodies against desmogleins and transmembrane cadherins which are essential for intercellular adhesion in desmosomes. Their disruption leads to loss of keratinocyte cohesion, resulting in the classic intraepidermal blistering.[8]

Early diagnosis based on histology and immunofluorescence findings is essential to initiate prompt treatment and prevent complications.[9] While the exact etiology of PV remains unclear, genetic susceptibility, particularly associations with HLA class II alleles, and environmental triggers such as medications (e.g., captopril, penicillamine), radiation, and infections have been implicated. [10]

Clinically, PV often begins with oral erosions in up to 50% of cases, which may precede cutaneous involvement. Cutaneous lesions typically develop on previously normal-appearing skin and evolve into raw, crusted erosions. Pruritus is uncommon, but persistent erosions may become secondarily infected.

Diagnosis is confirmed by a biopsy from the edge of an active lesion, with direct immunofluorescence demonstrating intercellular IgG and C3 deposits. When biopsy is not feasible, serological assays such as ELISA and indirect immunofluorescence are helpful alternatives. Before initiating treatment, baseline 2 of 3 laboratory tests and bone mineral density assessments are advisable due to the risk of corticosteroid-induced
adverse effects.[11]

The primary goal of therapy is to induce and maintain remission while minimizing drug-related complications. High-dose corticosteroids remain the first-line therapy, often combined with steroid-sparing immunosuppressants. The introduction of rituximab has significantly improved remission rates and is now favored as first-line treatment in
many guidelines. While untreated PV has a historically high mortality rate, timely and sustained immunosuppressive therapy has resulted in remission rates exceeding 75% within a decade of diagnosis.[12]

CONCLUSIONS

Pemphigus vulgaris is a rare but serious autoimmune blistering disease involving the skin and mucous membranes. Timely diagnosis and early initiation of effective treatment are essential to reduce morbidity and improve long-term outcomes. A multidisciplinary approach using targeted immunotherapy, such as rituximab, combined with corticosteroids and supportive care, offers the best chance for disease control and patient recovery. Prompt recognition and tailored treatment strategies are critical to ensuring favorable prognosis and preserving quality of life.

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