Clinicopathological Analysis of 27 Cases of Pemphigus from Jordan: A Retrospective Study

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ABSTRACT

Pemphigus is a serious blistering disorder with a rare presentations sometimes. Limited data exists on the clinicopathological features and treatment outcomes of pemphigus in Middle Eastern populations, particularly from Jordan.To analyze the clinical, histopathological features, treatment response and types of pemphigus in a group of our patients in Jordan. This retrospective cohort study analyzed 27 pemphigus cases diagnosed and treated between January 2015 and January 2025 across multiple medical centers in Jordan.Patient data included age, gender, pemphigus subtypes, clinical presentations, histopathological findings, treatment regimens, and therapeutic responses were collected and analyzed using SPSS version 25. Of the 27 patients (age range: 18-70 years, mean: 47.81 years), 19 (70.4%) were female. Pemphigus vulgaris was the predominant subtype (63%), followed by pemphigus foliaceus (29.6%), pemphigus erythematosus (3.7%), and pemphigus herpetiformis (3.7%). Mucocutaneous involvement was observed in 76% of pemphigus vulgaris cases. Histopathologically, suprabasal acantholysis characterized pemphigus vulgaris, whilepemphigus foliaceus cases revealed subcorneal/ intragranular acantholysis. Direct immunofluorescence was positive in 88.9% of cases. Patients receiving rituximab with prednisolone (18.5%) demonstrated superior clinical response with earlier steroid tapering compared to conventional immunosuppressive therapy. Diagnostic delays occurred in several cases, including two initially misdiagnosed as Behçet's disease and one pemphigus herpetiformis case undiagnosed for five years. This study highlights the clinical spectrum and treatment outcomes of pemphigus in a Jordanian population. Rituximab demonstrated strong efficacy and safety. Early recognition and accurate diagnosis are essential to optimize patient outcomes.

Keywords: Pemphigus vulgari, Pemphigus foliaceus, Acantholysis, Rituximab, Jordan

INTRODUCTION

Pemphigus is a rare group of autoimmune blistering disorders affecting the skin and mucous membranes, that histologically presents as intraepidermal blisters caused by acantholysis, and immunologically by IgG antibodies targeting desmogleins which are adhesion molecules essential for maintaining connections between keratinocytes[1]. The global incidence of pemphigus varies amongst different ethnic groups and geographical regions[2].

Pemphigus is classified into several subtypes, including pemphigus vulgaris, pemphigus foliaceus, pemphigus erythematosus, pemphigus vegetans, and pemphigus herpetiformis. These conditions are mediated by IgG autoantibodies that target desmosomal cadherins—desmoglein 3 in pemphigus vulgaris and vegetans, and desmoglein 1 in pemphigus foliaceus and pemphigus erythematosus , this results in disruption of keratinocyte adhesion and blister formation[3].

Pemphigus vulgaris is the most common severe variant, typically presents with flaccid bullae and painful erosions of skin and mucous membranes. Histologically, it is characterized by suprabasal acantholysis with a "tombstone" appearance of basal keratinocytes[4]. Pemphigus foliaceus is a more superficial form that presents with crusted erosions on seborrheic areas without mucosal involvement and demonstrates subcorneal acantholysis histologically[5]. Pemphigus erythematosus, considered a localized form of pemphigus foliaceus with features of lupus erythematosus which typically shows subcorneal acantholysis along with interface dermatitis[6].

Pemphigus vegetans is a rare variant of pemphigus vulgaris presenting clinically with vegetative plaques mainly on the flexures showing papillomatosis, pseudoepitheliomatous hyperplasia, and eosinophilic microabscesses histologically[7]. Pemphigus herpetiformis is a rare and atypical variant that mimics dermatitis herpetiformis clinically, with intensely pruritic, grouped vesicles and erythematous plaques and

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rarely if ever involves mucous membranes. It is reflected histologically by eosinophilic spongiosis and minimal or focal acantholysis, and direct immunofluorescence reveals intercellular IgG and/or C3 deposition, often directed against desmoglein 1[8].

The diagnosis of pemphigus relies on a combination of clinical features, histopathology, and immunofluorescence studies. Treatment typically involves systemic corticosteroids, often in combination with steroid-sparing immunosuppressive agents. In recent years, rituximab, which is a chimeric monoclonal antibody targeting CD20+ B cells, has emerged as an effective treatment option, particularly for refractory cases[9].

Despite advances in understanding the pathogenesis and management of pemphigus, data on the clinicopathological features and treatment outcomes in Middle Eastern populations, particularly from Jordan, remain limited. This study aims to provide a comprehensive analysis of the demographic patterns, clinical presentations, histopathological features, diagnostic challenges, and treatment responses across different pemphigus subtypes in a Jordanian cohort.

METHODS

Study Design and Participants: This retrospective cohort study analyzed all pemphigus cases diagnosed and treated by our team over a 10-year period from January 2015 to January 2025. Clinical records of 27 patients between the ages of 18 to 70 years were reviewed from multiple medical centers in Jordan. Inclusion criteria comprised patients diagnosed with one of the pemphigus subtypes who had comprehensive medical documentation available. Our cohort included individuals across diverse demographic backgrounds with data encompassing age, gender, pemphigus subtype, clinical presentation, histopathological findings, treatment regimens, and therapeutic responses.

Diagnostic Criteria: The diagnosis of pemphigus was established based on a combination of: 1. Clinical features: presence of characteristic skin and/or mucosal lesions 2. Histopathological findings: evidence of acantholysis at appropriate levels of the epidermis 3. Direct immunofluorescence (DIF): demonstration of intercellular IgG and/or C3 deposition in the epidermis. Pemphigus subtypes were classified according to standard clinical and histopathological criteria[10]. Pemphigus vulgaris was diagnosed based on suprabasal acantholysis and mucosal involvement, while pemphigus foliaceus was characterized by subcorneal/intragranular acantholysis without mucosal involvement. Pemphigus erythematosus was diagnosed when features of pemphigus foliaceus were accompanied by interface dermatitis, and pemphigus herpetiformis was identified by the presence of grouped vesicles, eosinophilic spongiosis, and minimal acantholysis.

Data Collection and Analysis: Patient data was collected using a standardized form that included demographic information, clinical features, histopathological findings, treatment regimens, and therapeutic responses. Clinical features were documented through detailed physical examinations and photographs . Histopathological analyses was performed on hematoxylin and eosin-stained sections of skin biopsies, and direct immunofluorescence studies were conducted on perilesional skin samples. Treatment regimens were grouped as follows: 1. Prednisolone combined with immunotherapy (azathioprine, mycophenolate mofetil, or cyclophosphamide) 2. Prednisolone combined with rituximab 3. Prednisolone monotherapy.

Therapeutic responses were evaluated based on clinical improvement, time to disease control, ability to taper corticosteroids, and relapse rates. Statistical analysis was performed using SPSS version 25. Descriptive statistics were calculated for demographic and clinical variables, including frequencies, percentages, means, medians, and standard deviations. Comparative analyses between treatment groups were conducted using appropriate statistical tests, with p-values of less than 0.05 considered statistically significant.

Ethical Considerations: This study was conducted in accordance with the Declaration of Helsinki and was approved by the Institutional Review Board of Mu'tah University Faculty of Medicine (approval number: MU-IRB-2025-37025). Patient confidentiality was maintained throughout the study and an informed consent was obtained from patients for the use of their clinical data and images for research and educational purposes.

RESULTS

Demographic Characteristics

A total of 27 patients with pemphigus were included in this study. The age range was 18-70 years, with a mean age of 47.81 years (median: 50 years, standard deviation: 15.30). The most frequent age was 60 years. Gender distribution showed a female predominance, with 19 females (70.4%) and 8 males (29.6%), with a female-to-male ratio of 2.4:1.

Distribution of Pemphigus Subtypes

Pemphigus vulgaris was the most common subtype, accounting for 17 cases (63.0%), followed by pemphigus foliaceus with 8 cases (29.6%). Pemphigus erythematosus and pemphigus herpetiformis were rare, each represented by a single case (3.7%). No cases of pemphigus vegetans were identified in our cohort (Figure 1).

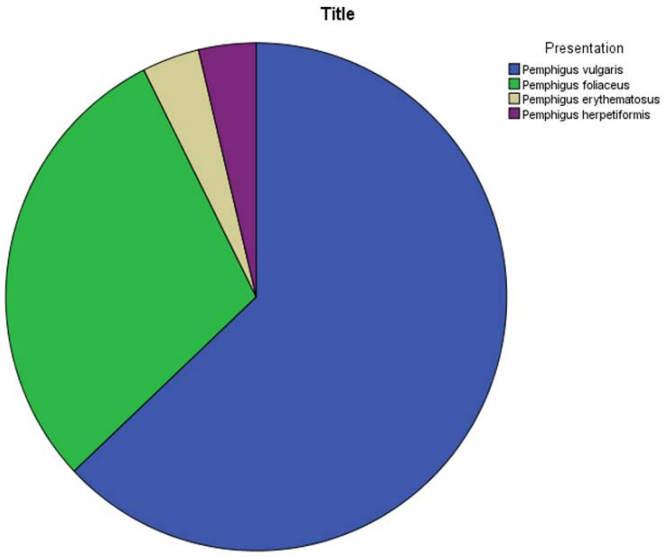

Figure 1. Distribution of pemphigus subtypes in the study cohort (n=27).

CLINICAL FEATURES

Pemphigus Vulgaris

Among the 17 patients with pemphigus vulgaris, 13 (76.5%) presented with mucocutaneous involvement, while 4 (23.5%) had isolated cutaneous lesions. The most common sites of involvement were the oral mucosa (76.5%), followed by the trunk (64.7%), scalp (47.1%), and extremities (41.2%). Notably, 2 patients (11.8%) presented with localized disease: one with an isolated facial ulcer persisting for more than 6 months, and another with localized scalp erosions and crusts of more than 6 months' duration (Table 1, Figures 2 and 3).

Table 1. Clinical Presentation of Pemphigus Vulgaris Cases (n=17)

Clinical Feature	Number of Patients	Percentage (%)
Mucocutaneous involvement	13	76.5
Cutaneous involvement only	4	23.5
Oral mucosal involvement	13	76.5
Trunk involvement	11	64.7
Scalp involvement	8	47.1
Extremities involvement	7	41.2
Localized disease	2	11.8

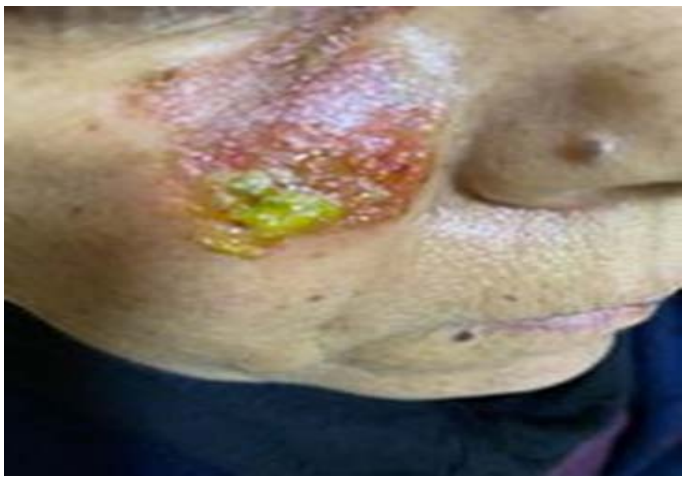

Figure 2. Localized single facial ulcer of more than 6 months duration (Our case with localized pemphigus vulgaris of the face).

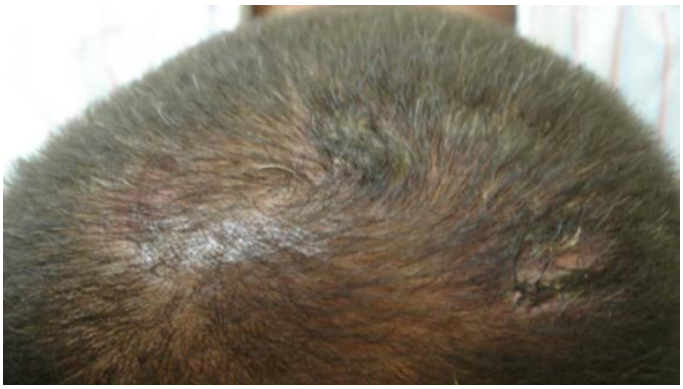

Figure 3. Localized scalp erosions and crusts of more than 6 months' duration.

Pemphigus Foliaceus

All 8 patients with pemphigus foliaceus presented with cutaneous involvement without mucosal lesions. The predominant sites of involvement were seborrheic areas, including the face (87.5%), scalp (75.0%), and upper trunk (62.5%). The clinical presentation typically consisted of crusted erosions and erythematous patches.

Pemphigus Erythematosus

The single case of pemphigus erythematosus presented with erythematous, scaly, and crusted lesions on the face in a butterfly distribution, resembling lupus erythematosus, along with scattered erosions on the upper trunk.

Pemphigus Herpetiformis

The patient with pemphigus herpetiformis presented with intensely pruritic grouped vesicular and crusted lesions on an erythematous base, primarily affecting the trunk and extremities. This patient had experienced symptoms for five years before the correct diagnosis was established, having been previously misdiagnosed with various conditions including allergic contact dermatitis and scabies. (Figure 4).

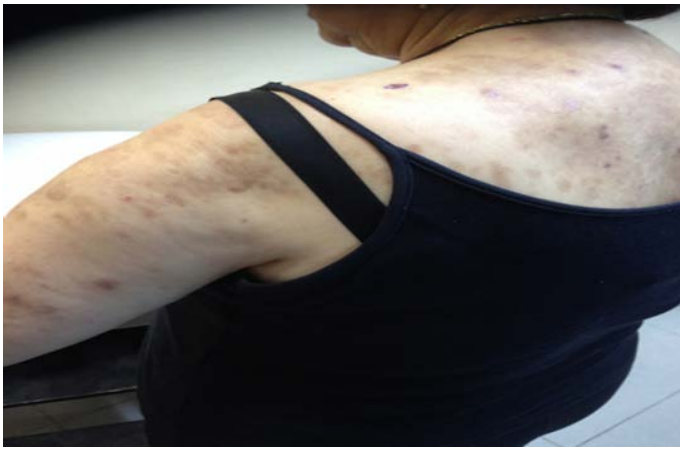

Figure 4. Multiple grouped vesicular and crusted lesions of 5 years duration (our case with pemphigus herpetiformis).

Histopathological Findings

The histopathological features of all cases were interpreted by a dermatopathologist .Acantholysis was observed in all pemphigus subtypes, with distinct patterns characterizing each of the variants (Table 2). In pemphigus vulgaris, acantholysis was typically suprabasal, with basal keratinocytes remaining attached to the basement membrane, creating the characteristic “row of tombstones” appearance (Figure 5). In pemphigus foliaceus, however , acantholysis was more superficial (intragranular), leading to a more superficial epidermal split. Pemphigus erythematosus and herpetiformis showed upper epidermal intragranular acantholysis (Figure-6), suggesting an overlap with pemphigus foliaceus.

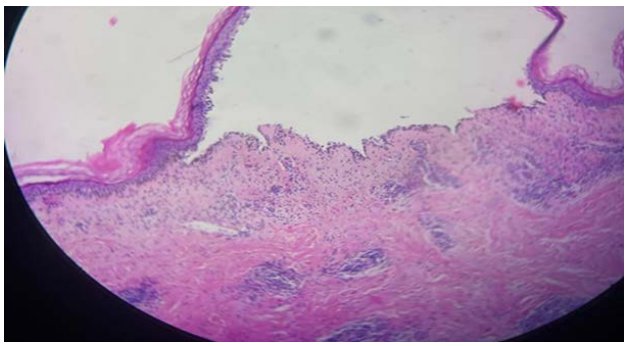

Figure 5. Suprabasal intraepidermal acantholytic split containing acantholytic cells with tombstone formation.

Dyskeratosis (abnormal keratinization), an occasional finding in pemphigus vulgaris reflecting epidermal response to inflammation and acantholysis[11], was present in one pemphigus vulgaris patient, three pemphigus foliaceus patients, and one pemphigus erythematosus case. Dense dermal inflammatory infiltrate, specifically of neutrophils and eosinophils, was observed in all cases of pemphigus vulgaris and foliaceus but was absent in the pemphigus erythematosus and herpetiformis cases.

Direct immunofluorescence (DIF) was positive in 24 of 27 cases (88.9%), showing intercellular IgG and/or C3 deposition in the epidermis. Specifically, DIF was positive in 15/17 (88.2%) pemphigus vulgaris cases, 7/8 (87.5%) pemphigus foliaceus cases, and in the single cases of pemphigus erythematosus and pemphigus herpetiformis.

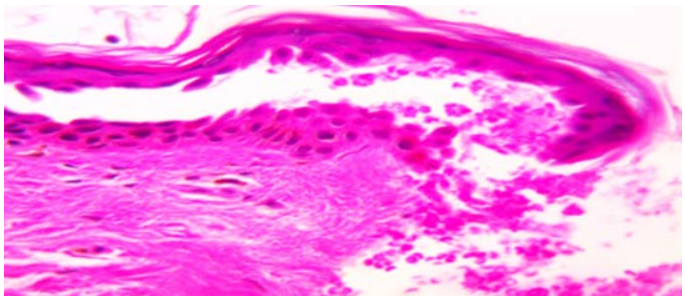

Figure 6. Upper intraepidermal acantholytic split with dyskeratosis containing acantholytic cells (a case of pemphigus herpetiformis)

Treatment Regimens and Responses

Various treatment regimens were employed based on disease severity and patient characteristics (Table 3). The majority of patients (22/27, 81.5%) received prednisolone in combination with conventional immunotherapy (azathioprine, mycophenolate mofetil, or cyclophosphamide). This group included 12 patients with pemphigus vulgaris, 8 with pemphigus foliaceus, 1 with pemphigus erythematosus, and 1 with pemphigus herpetiformis.

One patient with pemphigus vulgaris was treated with a combination of prednisolone and mycophenolate mofetil. A subgroup of 5 patients (18.5%) with pemphigus vulgaris received rituximab in combination with prednisolone. Notably, patients with pemphigus vulgaris who received rituximab showed excellent clinical response, which allowed for early tapering of systemic steroids and improved disease control compared to those on conventional therapy.

Table 2. Clinical & Histopathological Features of Pemphigus Subtypes

Feature	Pemphigus Vulgaris	Percentage %	Pemphigus Foliaceus	Percentage %	Pemphigus Erythematosus	Percentage %	Pemphigus Herpetiformis	Percentage %
Skin Involvement - Multiple Sites with mucous membrane involvement	13	48%	8	29%	1	4%	1	4%
Skin Involvement - Multiple Sites without mucous membrane involvement	1	4 %	8	100%	1	100%	1	100%
Skin Involvement only - Localized Site	2	7%	-		-		-	
Mucous Membrane Involvement without skin involvement	1	4%	-		-		-	
Acantholysis	17	63%	8	29%	1	4%	1	4%
Split Level	Suprabasal		Intragranular		Subcorneal / Intragranular		Subcorneal / Intragranular	
Dyskeratosis	1	4%	4	15%	1	4%	1	4%
Neutrophils & Eosinophils in Dermis	17	63%	8	29%	1	4%	1	4%
mDirect Imuno fluores cence Positive	15	-	7	-	1	-	1	-

Table 3. Treatment Regimens and Response

Treatment Regimen	Pemphigus Vulgaris (n=17)	Pemphigus Foliaceus (n=8)	Pemphigus Erythematosus (n=1)	Pemphigus Herpetiformis (n=1)	Total (n=27)
Prednisolone + Conventional Immunotherapy*	11 (64.7%)	8 (100%)	1 (100%)	1 (100%)	21 (77.8%)
Prednisolone + Mycophenolate Mofetil	1 (5.9%)	0 (0%)	0 (0%)	0 (0%)	1 (3.7%)
Prednisolone + Rituximab	5 (29.4%)	0 (0%)	0 (0%)	0 (0%)	5 (18.5%)
Time to disease control (weeks)†	8.2 ± 3.1 / 4.6 ± 1.8‡	6.4 ± 2.7	5.0	7.0	-
Steroid tapering (months)†	9.7 ± 3.8 / 5.2 ± 2.1‡	7.3 ± 2.9	6.0	8.0	-

*Conventional immunotherapy includes azathioprine or cyclophosphamide †Values presented as mean ± standard deviation ‡First value for conventional therapy / second value for rituximab therapy

Diagnostic Challenges

Several diagnostic challenges were encountered during the study period. Two cases were initially misdiagnosed as Behçet's disease, which delayed appropriate treatment by approximately one year. Additionally, the diagnosis of pemphigus herpetiformis was delayed by five years after symptom onset in one patient, highlighting the diagnostic difficulties associated with atypical presentations of pemphigus.

DISCUSSION

This retrospective clinicopathological study of 27 pemphigus patients from multiple medical centers in Jordan provides a comprehensive overview of the demographic patterns, clinical manifestations, histopathological features, diagnostic challenges, and therapeutic responses associated with different subtypes of pemphigus. To our knowledge, this represents one of the few studies examining pemphigus in the Jordanian population, offering valuable insights into the disease characteristics in this region.

Demographic and Clinical Patterns

The predominance of pemphigus vulgaris (63%) in our cohort aligns with global epidemiological trends, where it is consistently reported as the most common and severe form of the disease[12]. This reinforces the fact that pemphigus vulgaris remains the primary burden among other autoimmune blistering disorders both regionally and internationally. The female predominance (70.4%) observed in our study is consistent with previous literature, where autoimmune disorders, including pemphigus, more frequently affects women[13,14]. The mean age of onset (47.81 years) also corresponds with international data suggesting a peak incidence in the fourth to sixth decades of life[14]. These demographic patterns suggest that genetic and environmental factors influencing pemphigus susceptibility may be similar across different populations.

The mucocutaneous involvement in 76.5% of pemphigus vulgaris cases highlights its characteristic presentation and the importance of recognizing both cutaneous and mucosal lesions in clinical evaluations[15]. Interestingly, we observed isolated presentations involving the scalp, face, or mucosa in some patients, reflecting the spectrum of disease manifestations and emphasizing how important it is to consider pemphigus in atypical erosive or ulcerative lesions, particularly in chronic or treatment-resistant cases.

Histopathological Correlations

Histopathological examination revealed acantholysis in all subtypes, with distinct split levels aiding differentiation: suprabasal in pemphigus vulgaris and subcorneal/intragranular in pemphigus foliaceus. These findings are consistent with established histopathological criteria for pemphigus subtypes[16]. The presence of dermal eosinophils and neutrophils, particularly in pemphigus vulgaris and foliaceus, supports an active inflammatory component, which may influence therapeutic response and disease progression[17]. Direct immunofluorescence positivity in 88.9% of cases supports its role as a gold standard for diagnosis[18]. The small percentage of DIF-negative cases (11.1%) may be attributed to technical factors, sampling errors, or the effect of prior treatments, as reported in other studies.

Diagnostic Challenges and Implications

A significant finding in our study was the diagnostic delay in several cases. Two patients were initially misdiagnosed with Behçet's disease, and one case of pemphigus herpetiformis remained undiagnosed for five years. These diagnostic pitfalls highlight the need for improved clinician awareness and broader consideration of pemphigus variants in the differential diagnosis of erosive mucocutaneous disorders. The overlap in clinical and histopathological features, particularly between pemphigus foliaceus and erythematosus, and the atypical presentation of pemphigus herpetiformis, further complicates diagnosis without immunofluorescence support. Our experience suggests that a high index of suspicion and early referral for specialized dermatological evaluation and appropriate immunodiagnostic testing are crucial for timely diagnosis and management.

Treatment Outcomes and Therapeutic Implications

Therapeutically, our findings suggest that rituximab, a CD20 monoclonal antibody, showed superior efficacy in pemphigus vulgaris patients. Those treated with rituximab exhibited a faster steroid tapering timeline and improved clinical control, supporting its role as a potential first-line treatment in moderate to severe disease. This mirrors findings from international clinical trials and growing consensus among dermatologists advocating for early biologic intervention to minimize long-term steroid exposure and related side effects[19,20]. Traditional immunosuppressants combined with corticosteroids was also effective in many cases but may not provide the rapid control or reduced adverse effect profile offered by rituximab[21]. The selection of the most appropriate therapy should be individualized based on disease severity, patient comorbidities, and access to biologics, which remains a challenge in many healthcare settings, particularly those in developing countries[22].

Study Limitations: There were several limitations to be considered in our study . Of these limitations is the small sample size and the retrospective design which limit statistical power and may introduce selection bias. The lack of long-term follow-up data on relapse rates and treatment durability, especially for rituximab-treated patients, prevents definitive conclusions about long-term outcomes. Furthermore, we did not perform serological studies such as ELISA for anti-desmoglein antibodies, which could have provided additional diagnostic and prognostic information. Future prospective studies with larger cohorts, standardized diagnostic protocols, and longer follow-up periods would address these limitations and provide more robust evidence for optimizing pemphigus management in the Middle Eastern population.

CONCLUSION

This study provides valuable insights on the clinicopathological features and treatment outcomes of pemphigus in a Jordanian population. Our findings highlight several key aspects of this rare autoimmune blistering disorder in the Middle Eastern context. Pemphigus vulgaris emerged as the predominant subtype, affecting primarily middle-aged females, with characteristic mucocutaneous involvement in the majority of cases. Histopathological examination confirmed distinct acantholytic patterns across different subtypes, with direct immunofluorescence serving as a reliable diagnostic tool in nearly 90% of cases. The diagnostic challenges encountered in our cohort, including misdiagnosis and delayed diagnosis in several cases, underscore the importance of increased awareness among clinicians about the varied presentations of pemphigus and the need for early referral for specialized dermatological evaluation and appropriate immunodiagnostic testing. From a therapeutic perspective, rituximab demonstrated superior efficacy and safety in patients with moderate to severe pemphigus vulgaris, allowing for earlier steroid tapering and improved disease control compared to conventional immunosuppressive therapy. This finding supports the growing international consensus regarding the role of rituximab as a frontline therapy in pemphigus management. The limitations of our study, including its retrospective nature,

small sample size, and lack of long-term follow-up data, highlight the need for larger prospective studies with standardized protocols and extended follow-up periods to further validate our findings and optimize pemphigus management in Middle Eastern populations. In conclusion, early recognition, accurate diagnosis, and appropriate treatment selection are essential for optimizing outcomes in patients with pemphigus. Our study contributes to the limited literature on pemphigus in the Middle East and provides a foundation for future research in this field.

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Competing Interest: None

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