DERMATOLOGY

INDEX COPERNICUS INTERNATIONAL

ICV value for 2024 = 82.51

Case Report | International Journal of Medical Case Reports

Pemphigus Vulgaris with Extensive Mucocutaneous Involvement: Importance of Early Recognition of Oral Lesions – A Case Report.

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ISSN 2455-0574

ABSTRACT

Background:

Pemphigus vulgaris (PV) is a chronic autoimmune blistering disorder usually presenting in 5th decade of life. The hallmark feature of PV is intraepidermal acantholysis which is mediated by autoantibodies against desmoglein 1 and 3. PV predominantly affects mucous membranes and skin. Many cases often present initially only with oral lesions after which cutaneous involvement is gradually noticed. Early diagnosis is critical from the point of view of managing these patients as delayed recognition can lead to significant morbidity and mortality due to complications. In patients of PV oral manifestations are frequently misdiagnosed initially, thereby resulting in delayed treatment. This report highlights the importance of early identification of oral lesions in cases of PV and underscores the need of multidisciplinary approach in these cases.

Case Report:

A 35-year-old female presented with 1 month history of painful oral ulcers. These ulcers initially appeared as fluid-filled blisters that later ruptured to form erosions. There was a history of similar lesions gradually developing over the chest and abdomen. Clinical examination revealed multiple oral erosions involving the buccal mucosa and palate along with flaccid bullae and erosions on the trunk. Nikolsky's sign was positive. Tzanck smear demonstrated acantholytic cells. Histopathological examination showed suprabasal clefting with a "row of tombstone" appearance of basal keratinocytes. Clinical features and histopathology confirmed the diagnosis of PV. The patient was treated with systemic prednisolone and azathioprine therapy along with topical therapy as well as supportive care. Significant clinical improvement occurred within two weeks of starting therapy with reduced lesion formation and improved oral intake.

Conclusion:

This case underscores the importance of identifying oral manifestations of PV and doing further evaluation in selected cases. Early recognition and timely initiation of immunosuppressive therapy are important in preventing disease progression and improving outcomes. A high index of suspicion for possibility of PV in cases with oral ulcers is essential to reduce diagnostic delays.

Keywords: Acantholysis, Autoimmune Diseases, Histopathology, Immunosuppressive Therapy, Pemphigus Vulgaris.

INTRODUCTION

Pemphigus vulgaris (PV) is a potentially life-threatening autoimmune blistering disorder. It is characterized by loss of keratinocyte adhesion resulting in fragile intraepithelial bullae and painful erosions involving the skin and mucous membranes . Although PV is uncommon in the general population, it carries substantial clinical significance because of its chronic relapsing course, the severity of mucocutaneous involvement, and the morbidity associated with delayed diagnosis and treatment . Epidemiologically, PV most often affects adults in the fifth and sixth decades of life, with geographic and ethnic variation in incidence, and a slight female predominance reported in several populations. However, younger adults may also be affected, and such presentations can be diagnostically challenging because clinicians may not initially suspect an autoimmune vesiculobullous disorder in this age group. The disease burden extends beyond visible lesions, as persistent pain, dehydration, weight loss, secondary infection and psychosocial distress markedly reduce quality of life. Therefore, recognition of PV at an early stage is essential to prevent progression from localized mucosal disease to extensive mucocutaneous involvement.[1]

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International Journal Of Medical Case Reports (ISSN 2455-0574) is an indexed medical journal indexed in Index Copernicus

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In PV autoantibodies directed against Dsg3 predominantly result in mucosal involvement whereas antibodies against both Dsg1 and Dsg3 lead to mucocutaneous manifestations. Binding of these autoantibodies is known to disrupt keratinocyte adhesion which results in loss of intercellular cohesion and blister formation within the epidermis. Histopathologically, PV is characterized by suprabasal clefting and presence of acantholytic (Tzanck) cells. Additionally, a distinctive "row of tombstone" appearance of basal keratinocytes is also highly suggestive of PV. Direct immunofluorescence (DIF) remains the gold standard for diagnosis which demonstrates intercellular deposition of IgG and C3 in a characteristic fish-net pattern. These histopathological features are important for distinguishing PV from cases of other similar vesiculobullous disorders.[2]

Clinically, PV most commonly presents with painful erosions of the oral mucosa. These erosions often precede cutaneous involvement by many weeks to months. Oral lesions are reported as the initial manifestation in majority of the cases and in many cases, they may remain the sole presentation for a prolonged period. These lesions typically involve the buccal mucosa, palate and gingiva. The lesions in PV usually present as fragile bullae that rupture easily leaving behind raw and erythematous erosions. Cutaneous lesions usually appear as flaccid bullae on normal or erythematous skin which rupture to form characterstic crusted erosions. A positive Nikolsky's sign is the hallmark clinical feature of PV. The involvement of oral mucosa significantly impairs nutrition and quality of life thereby in many cases leading patients to seek dental consultation before dermatological evaluation. Early recognition by dental and primary care physicians plays an important role in reducing delay in diagnosis and preventing disease progression.[3]

The management of PV has evolved significantly over the past decades. In majority of the cases systemic corticosteroids remaining the cornerstone of therapy. Adjunctive immunosuppressive agents such as azathioprine, mycophenolate mofetil and cyclophosphamide are commonly used as steroid-sparing agents to minimize long-term adverse effects in patients. More recently, biologic therapies such as rituximab have emerged as highly effective therapeutic option. Rituximab targets CD20-positive B cells thereby reducing autoantibody production. Rituximab therapy started early in the course of illness has been shown to improve prognosis, reduce disease severity and decrease mortality rates. [4]

This case of PV highlights the importance of recognizing early mucosal manifestations and correlating clinical findings with histopathological and immunofluorescence studies. The present case report aims to contribute to the existing literature by emphasizing the diagnostic significance of oral lesions in cases of PV and to reinforce the need for timely multidisciplinary intervention to improve patient outcomes.

CASE REPORT

A 35-year-old female presented to the dermatology outpatient department with complaint of painful oral ulcers since one-month. TO begin with lesions appeared as fluid-filled blisters that ruptured spontaneously and left behind painful erosions. Over time, similar lesions developed over the chest and abdomen. The patient reported a burning sensation in the oral cavity which was aggravated by spicy food along with significant difficulty in eating. There was associated pruritus and watery discharge from the cutaneous lesions. She also complained of photosensitivity with exacerbation of lesions upon sun exposure. There was no history of fever, weight loss, recent drug intake or any previous similar episodes.

On general examination the patient was conscious, and well -oriented to time, place, and person with stable vital signs. Intraoral examination revealed multiple irregular erosions involving the buccal mucosa and palate. These lesions were found to have with desquamative areas over an erythematous base. Nikolsky's sign was positive. Further general examination demonstrated multiple flaccid bullae and erosions over the chest and abdomen with areas of raw, oozing surfaces following rupture of blisters. Systemic examination did not reveal any abnormalities. (Figure 1, Figure 2)

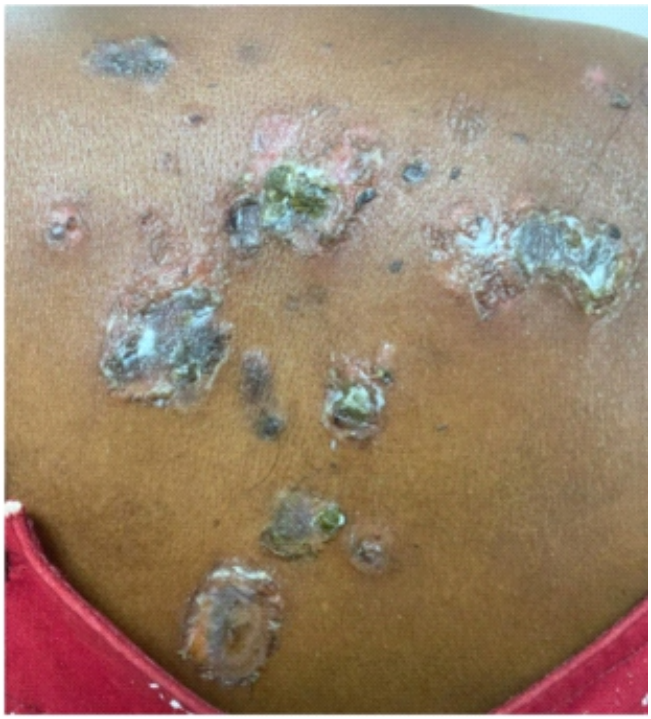

Figure 1:- Clinical image showing multiple flaccid bullae and ruptured vesiculobullous lesions over the anterior abdominal wall, with irregular erosions, crusting, and areas of raw oozing surfaces—features consistent with cutaneous involvement in pemphigus vulgaris.

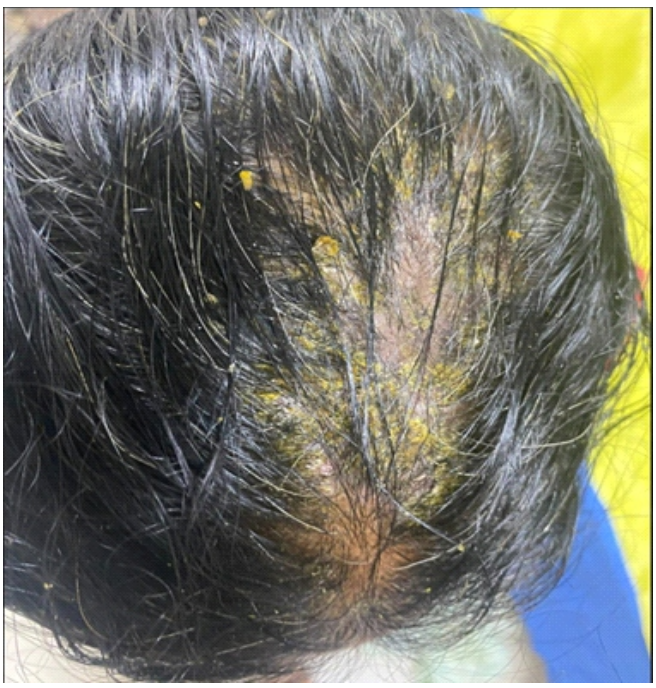

Figure 2: Scalp showing erosive lesions with crusting and scaling over an erythematous base, along with patchy hair involvement—findings suggestive of scalp involvement in pemphigus vulgaris.

Routine laboratory investigations, including complete blood count, were within normal limits, while erythrocyte sedimentation rate was mildly elevated. Acantholytic cells were seen on Tzanck smear. Histopathological examination showed suprabasal cleft formation with acantholytic cells and a characteristic row of tombstone basal keratinocytes. Based on the clinical presentation and histopathological findings a definitive diagnosis of pemphigus vulgaris was established .

The patient was initiated on systemic therapy with oral prednisolone 60 mg once daily and azathioprine 50 mg twice daily. Additionally topical clobetasol propionate 0.05% was also prescribed for cutaneous lesions over chest and abdomen. 2% lidocaine oral gel was given for local application for symptomatic relief of oral lesions. Supportive care and counseling were provided, and the patient was advised regular follow-up with monitoring for potential adverse effects of immunosuppressive and systemic corticosteroid therapy.

At two-week follow-up visit the patient reported significant clinical improvement. There was a marked reduction in the formation of new blisters. Also, there was progressive healing of existing oral as well as cutaneous lesions. She reported significant pain relief and considerably improved ability to tolerate oral intake. A gradual tapering of corticosteroids was planned depending upon clinical response. Patient was advised continued monitoring for appearance of new lesions and to report immediately is any treatment-related complications appear.

DISCUSSION

The present case is notable because painful oral erosions were the earliest and most functionally disabling manifestation, with subsequent progression to truncal bullae and erosions. This pattern is highly consistent with the clinical observations reported by Esmaili et al[5] and Shamim et al[6] both of whom emphasized that the oral cavity is a common initial site of disease activity in pemphigus vulgaris (PV). Our patient showed the same topographic pattern, with erosions involving the buccal mucosa and palate, followed by cutaneous lesions over the chest and abdomen. Although PV classically affects patients in the fifth to sixth decades, Shamim et al reported a mean age of 42.73 years, and our patient was even younger at 35 years, underscoring that younger adults can also develop clinically significant PV and may therefore be at greater risk of delayed recognition when autoimmune blistering disease is not initially suspected. The severe pain, burning sensation, and difficulty in oral intake in our patient also mirror the substantial nutritional and quality-of-life burden described in oral-predominant PV.

The mucocutaneous progression observed in this patient is also reported by many other authors. Amagai et al identified the pemphigus vulgaris antigen as a novel epithelial cadherin, now recognized as desmoglein 3, establishing the molecular basis for acantholysis in PV.[7] Subsequently, Salato et al showed that mucosal PV is associated primarily with autoimmunity against desmoglein 3, whereas transition to mucocutaneous disease is associated with broader autoreactivity involving both desmoglein 3 and desmoglein 1 through epitope spreading.[8] This framework explains the chronology in the present patient: the disease began with oral blisters and erosions, then extended to the skin of the trunk, suggesting evolution from a mucosal-dominant phase to mucocutaneous PV. The case therefore provides a clinicopathologic illustration of the desmoglein compensation concept in routine practice. It also reinforces an important diagnostic message: oral lesions in PV are not merely localized manifestations but may represent the first clinically visible stage of a more disseminated autoimmune process. Recognizing oral PV early is therefore essential not only for symptom relief but also for interrupting progression to more extensive cutaneous involvement, secondary infection, fluid loss, and treatment delay. In this sense, our patient's presentation strongly supports the concept that oral erosions should be interpreted as a sentinel sign of potentially evolving systemic disease rather than as an isolated oral disorder.

The diagnostic findings in the present case were classic and internally concordant, with a positive Tzanck smear, suprabasal clefting with acantholytic cells and "row of tombstones" on histopathology. These results are in line with the observations of Coscia-Porrazzi et al who demonstrated the utility of cytology in oral PV, showing that cytomorphology and DIF-based assessment of oral smears can aid early diagnosis in suspected oral pemphigus.[9] The current case also highlights the stepwise value of combining bedside, histologic, and immunopathologic tests. Tzanck smear in our patient provided rapid preliminary evidence by demonstrating acantholytic cells, which was particularly useful given the fragile mucosa and active erosions. However, as prior studies have shown, cytology alone is supportive rather than definitive; confirmation still depends on tissue histopathology and immunofluorescence. The present case therefore reaffirms the enduring diagnostic importance of perilesional biopsy and histopathology in suspected PV. In settings where oral lesions are initially assessed by dentists, oral physicians, or primary-care clinicians, early referral for biopsy and DIF can substantially shorten the diagnostic interval. This is especially relevant because oral PV may clinically resemble aphthous ulceration, erosive lichen planus, candidiasis, erythema multiforme, or other ulcerative disorders, leading to potentially avoidable misclassification and delayed treatment.

The favorable early therapeutic response in our patient after initiation of systemic prednisolone along with azathioprine is consistent with many studies. Beissert et al compared oral methylprednisolone plus azathioprine with methyl-prednisolone plus mycophenolate mofetil and showed that azathioprine-containing regimens could achieve complete remission in a substantial proportion of patients.[10] Our patient experienced marked reduction in new blister formation and progressive healing of both oral and cutaneous lesions within two weeks, together with improved oral intake, which is clinically meaningful because restoration of feeding is often one of the earliest indicators of treatment success in oral-predominant PV. The quick symptomatic relief observed here also underscores the importance of adjunctive local care, including topical corticosteroids for skin lesions and topical anesthetic therapy for painful oral erosions. Although a two-week response does not establish long-term remission, it strongly suggests good early disease control and supports the appropriateness of corticosteroid-based induction combined with azathioprine in this case. At the same time, careful follow-up remains essential because relapse, steroid toxicity, azathioprine-related adverse effects, and secondary infection remain important concerns during the chronic course of PV.

Finally, the present case highlights the broader clinical lesson that early recognition of oral PV should trigger multidisciplinary management and may reduce the need for prolonged high-dose corticosteroid exposure. In the Ritux 3 trial, Joly et al[11] showed that first-line rituximab combined with short-term prednisone was more effective than prednisone alone and was associated with fewer adverse events, while Murrell et al [12] and the international panel formalized contemporary recommendations for diagnosis and management of pemphigus, emphasizing standardized assessment and timely escalation of therapy when indicated.

Although our patient responded well to prednisolone and azathioprine, these newer data are still relevant because they frame current expectations for disease control and steroid minimization, particularly in moderate-to-severe disease or in patients with relapse. More importantly for this case report, both studies indirectly reinforce the value of early diagnosis: the earlier PV is identified, the earlier effective immunosuppressive therapy can be started, and the greater the opportunity to limit disease extension, cumulative steroid burden, hospitalization, malnutrition, and psychosocial distress. Thus, the chief message from this case is not only that oral lesions may precede skin lesions, but that they often provide the best window for early intervention. Dentists, dermatologists, pathologists, and primary-care physicians must therefore maintain a high index of suspicion for PV in any patient with persistent, painful, nonhealing oral erosions, particularly when lesions are preceded by fragile blisters or accompanied by a positive Nikolsky sign.

CONCLUSION

Pemphigus vulgaris may initially present with isolated oral erosions before progressing to widespread mucocutaneous lesion. This is more so in younger patients where clinical suspicion may be low. Early recognition of characteristic oral lesions supported by timely histopathological evaluation is important for early diagnosis and timely initiation of appropriate therapy. Starting immunosuppressive therapy at an early stage is known to significantly reduce disease progression and improve patient outcomes.

Conflict Of Interest : None

Source Of Funding : None

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Author Contribution :
KS-contributed to patient management, data collection, and manuscript drafting. **VK**-participated in data interpretation, literature review, and critical manuscript revision.

Received : 01-02-2026 Revised: 25-02-2026 Accepted : 15-03-2026