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Pemphigus Vulgaris Presenting as Severe Crusted Lesions of the Lower Lip: A Case Report

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Abstract: Pemphigus vulgaris (PV) is a rare autoimmune blistering disease of the skin and mucous membranes. The oral cavity is frequently the first, and sometimes the only, site involved, and crusted or erosive lesions may be mistaken for malignancy. A 70-year-old woman in good general health was referred for crusted lesions on an erythematous background involving the entire lower lip and its inner surface, highly suspicious of malignancy. An erosive plaque of the right buccal mucosa, measuring 2 cm along its longest axis, with raised borders and surrounding erythema, had been evolving for 3 months. No other cutaneous site was involved, and the Nikolsky sign was positive. Biopsies of two lesions were performed. Histopathology and direct immunofluorescence (DIF) were consistent with pemphigus vulgaris. The patient received local prednisone 20 mg daily together with topical treatment of the labial lesions. Marked clinical improvement was observed after 2 months of treatment. Lessons. Severe crusted lip lesions in an older adult can present as PV rather than carcinoma. Early biopsy with DIF, and attention to a positive Nikolsky sign, allow prompt diagnosis and treatment of an otherwise misleading presentation.

Keywords: *Pemphigus Vulgaris; Oral Pemphigus; Lip Lesions; Oral Squamous Cell Carcinoma; Differ; Corticosteroids; Case Report.*

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I. INTRODUCTION

Pemphigus vulgaris (PV) is a rare, potentially life-threatening autoimmune blistering disease caused by IgG autoantibodies directed against desmosomal cadherins, mainly desmoglein 3 (Dsg3) and, in mucocutaneous disease, desmoglein 1 (Dsg1). These antibodies disrupt keratinocyte adhesion (acantholysis) and produce suprabasal intraepithelial blisters [1-4]. Incidence varies with geographic and ethnic background, with higher rates reported in some Mediterranean and Middle Eastern populations, and onset typically occurs in middle age or later [1,2].

Oral involvement is the rule rather than the exception. The oral mucosa is affected in most patients, is often the presenting site, and may remain the only site involved for months [6-8,10]. Blisters rupture rapidly in the mouth, so patients usually present with painful, irregular, slowly healing erosions of the buccal mucosa, palate or gingiva, and less often of the lips [6-9]. Because these erosions are chronic and may be crusted or indurated, they can be confused with erosive lichen planus, mucous membrane pemphigoid, erythema multiforme, paraneoplastic pemphigus, or squamous cell carcinoma (SCC) [6,8,10-12].

The lower lip is a particularly concerning site in an older patient because lip SCC is common at this location and because hemorrhagic, crusted ulceration raises concern for malignancy. Misdiagnosis can lead to unnecessary surgery or delay appropriate immunosuppressive treatment. We report a severe presentation of oral PV, with crusted involvement of the entire lower lip and an erosive buccal plaque, referred for suspected malignancy.

II. CASE PRESENTATION

A 70-year-old woman in good general health was referred to the Department of Dental Medicine at Taher Sfar Teaching Hospital for crusted lower-lip lesions highly suspicious for malignancy. The lesions covered the entire lower lip and its inner surface. A separate lesion of the right buccal mucosa had been evolving for 3 months. The labial lesions had a rapid onset and progression.

The patient had no relevant medical or surgical history or family history of autoimmune disease. There was no reported medication use, dysphagia, or weight loss. No tobacco or alcohol consumption or sun exposure was identified.

Extraoral examination showed crusted lesions on an erythematous background involving the whole lower lip. The crusts were thick, dark, and hemorrhagic, extending onto the inner lip surface (Figures 1 and 2). No regional lymphadenopathy or other cutaneous involvement was noted. Intraoral examination showed an ulcerative plaque of the right buccal mucosa with raised borders and surrounding erythematous mucosa, measuring 2 cm along its longest axis

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(Figure 3). The Nikolsky sign was positive. The oral hygiene was poor.



Fig 1 Extraoral View Showing the Labial Lesions



Fig 2 Extraoral View Showing Lesions on the Inner Surface of the Lower Lip



Fig 3 Intraoral Photograph Showing an Inhomogeneous Ulcerative Lesion of the Right Buccal Mucosa.

Table 1 Timeline

Time point	Event
About 3 months before presentation	Onset of the right buccal erosive plaque; onset of labial lesions [X]
Week 1	Referral and consultation: crusted lesions of the entire lower lip, suspected malignancy; positive Nikolsky sign
Week 1 (same session)	Biopsies of two lesions
Week 2	Histopathology and DIF consistent with pemphigus vulgaris
Week 2	Start of prednisone 20 mg daily and topical treatment of the labial lesions
2 months after treatment start	Marked clinical improvement

The hemorrhagic, crusted lower lip in a 70-year-old patient first raised the suspicion of squamous cell carcinoma (SCC). Two lesions were therefore biopsied. Histopathological examination revealed stratified squamous epithelium with suprabasal acantholysis and intraepithelial cleft formation. The basal cell layer remained attached to the basement membrane, exhibiting a characteristic “tombstone” appearance. Detached acantholytic keratinocytes (Tzanck cells) were observed within the cleft. The underlying connective tissue showed a mild chronic inflammatory infiltrate composed mainly of lymphocytes and plasma cells. Direct immunofluorescence examination of perilesional oral mucosa demonstrated intercellular deposition of IgG and C3 within the epithelium, in a characteristic “fishnet” pattern. These findings confirmed the diagnosis of pemphigus vulgaris.

The patient was treated with local prednisone 20 mg daily. The patient received this dose 3 times per day for 3 weeks, then 2 times per day for 10 days, then once daily for another 10 days. Topical treatment of the labial lesions consisted of Clobetasol applied 3 times per day for one month, then once daily for another month. Adjuvant measures included improved oral hygiene, scaling, and antiseptic mouthwash.

At the 2-month follow-up, the patient showed marked clinical improvement. The labial crusts and erosions substantially decreased, buccal plaque regressed, and the Nikolsky sign was no longer elicited. The lesions showed partial healing, with a marked reduction in lesion extent (Figures 4 and 5). No treatment-related adverse events were reported. Patient-reported outcomes also improved, with reduction in pain and improved ability to eat. Regular follow-up is planned, including clinical assessment of disease activity using the Pemphigus Disease Area Index (PDAI).

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Fig 4 Reduction in Lesion Extent at 2 Months Follow-Up



Fig 5 Marked Clinical Improvement of the Buccal Lesion at 2 Months Follow-Up

III. DISCUSSIONS

PV results from IgG autoantibodies against Dsg3 and, in mucocutaneous disease, Dsg1 [4]. According to the desmoglein compensation hypothesis, the autoantibody profile determines the clinical phenotype: anti-Dsg3 antibodies alone are associated with mucosal-dominant disease, whereas the addition of anti-Dsg1 antibodies produces skin involvement [5]. The strictly oral and labial distribution in this patient is compatible with a mucosal-dominant profile. A positive Nikolsky sign, which reflects loss of keratinocyte cohesion, may be elicited on perilesional or apparently normal epithelium and supports an acantholytic disease [2,3].

The oral mucosa is frequently the initial site of PV, and mucosal-dominant PV can remain limited to the mouth for a prolonged period [6-8]. Erosions usually occur on the buccal mucosa, palate, and gingiva, whereas lip involvement is less commonly emphasized [6-9]. Hemorrhagic crusting of the lips is classically associated with erythema multiforme major, Stevens-Johnson syndrome, and paraneoplastic pemphigus [10,11]. The extensive crusting of the entire lower lip in this patient therefore illustrates an unusually severe presentation of oral PV and explains the initial suspicion of malignancy.

Several pitfalls can delay diagnosis. First, a chronic, crusted lip lesion in an older person is appropriately managed as a possible SCC, but a biopsy from a necrotic or crusted area may show only nonspecific inflammation. Biopsy of the margin of an erosion, including adjacent intact mucosa, is preferable [10,14]. Second, DIF requires a perilesional specimen handled in an appropriate transport medium, not formalin, and histology alone cannot distinguish PV from other erosive diseases [10,14]. Third, erosive lichen planus and mucous membrane pemphigoid share a clinical appearance with oral PV and are distinguished by histopathology and DIF [6,12]. Finally, paraneoplastic pemphigus must be considered when mucosal disease is severe and hemorrhagic, because it is associated with an underlying neoplasm and a poorer prognosis [11]. Coexistence of PV and a true malignancy is possible, and a negative carcinoma biopsy should be interpreted in light of the clinical course.

Systemic corticosteroids remain the cornerstone of initial treatment. Guidelines usually describe prednisolone at about 1 mg/kg/day in moderate to severe disease [13-15]. The dose used here, 20 mg daily, is lower than such weight-based regimens; the rationale, for example, disease limited to the oral mucosa or comorbidity, should be stated by the authors [X]. Topical corticosteroids are used as adjuncts for oral and labial lesions [6,13]. In extensive or refractory disease, or when corticosteroid toxicity is a concern, current guidelines recommend rituximab, and a randomized trial showed that first-line rituximab with short-term prednisone achieved more frequent sustained remission off therapy than prednisone alone [15,16]. Rituximab was also superior to mycophenolate mofetil in sustained steroid-free remission [17]. Azathioprine and mycophenolate mofetil are used as steroid-sparing adjuvants, with more modest evidence [13,14,19]. In our patient, the marked improvement after 1 month is consistent with the usual topical corticosteroid responsiveness of PV, but oral lesions may be slow to clear, and relapses are common during tapering [6,18].

Prognosis has improved considerably since the introduction of corticosteroids and immunosuppressants, but PV remains a chronic, relapsing disease with a mortality that is largely related to infection and treatment complications [1,2,18]. Long-term follow-up is therefore required. This report has limitations: it describes a single patient with a follow-up of 2 months. Its strengths are the striking clinical presentation and the confirmation by histopathology and DIF.

IV. CONCLUSION

Oral PV can present as severe crusted lesions of the entire lip that closely mimic carcinoma. Key lessons:

- A crusted or erosive lip or buccal lesion with a positive Nikolsky sign should prompt consideration of an autoimmune bullous disease.

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- Biopsy for histopathology and DIF should be obtained early, from the margin of the lesion and perilesional mucosa.
- Topical corticosteroids can produce rapid improvement, and long-term monitoring, a careful taper, and early recourse to steroid-sparing therapy in refractory disease are necessary.

INFORMED CONSENT AND DECLARATIONS

Written informed consent for publication of this case report and the accompanying clinical photographs was obtained from the patient.

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