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Mucosal involvement in pemphigus foliaceus: A rare presentation

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Abstract
Pemphigus foliaceus (PF) is a rare autoimmune blistering disorder characterized by antibodies targeting desmoglein 1 (Dsg1), a critical adhesion protein in the upper epidermis. Mucosal involvement is typically absent due to the protective expression of desmoglein 3 (Dsg3) in mucous membranes. However, rare cases challenge this paradigm, indicating the potential for atypical presentations. We report a 62-year-old woman with type 2 diabetes who presented with chronic, painful erosions of the oral mucosa, followed by development of erosive cutaneous lesions. Histopathology and direct immunofluorescence confirmed the diagnosis of PF, and ELISA identified circulating anti-Dsg1 antibodies in the absence of anti-Dsg3. Treatment with systemic corticosteroids and rituximab led to rapid skin improvement, though a subsequent relapse of isolated oral lesions occurred. Colchicine therapy provided significant and sustained relief. This case highlights the rare but significant potential for mucosal involvement in PF, possibly due to high autoantibody titers or chronic inflammation compromising mucosal integrity. It underscores the need for clinicians to include PF in the differential diagnosis of persistent oral erosions and to consider adjunctive treatments like colchicine for refractory mucosal disease. A tailored, multimodal approach can effectively manage both cutaneous and mucosal manifestations in such atypical presentations.

Keywords: Pemphigus foliaceus, oral mucosa, desmoglein 1, mucosal involvement, autoimmune blistering disease

Introduction
Pemphigus foliaceus, also known as seborrheic pemphigus, is a rare autoimmune blistering disease caused by autoantibodies targeting desmoglein 1 (Dsg1), a cadherin-type protein involved in epidermal cohesion. The disease typically affects the skin and classically spares mucous membranes due to the predominant expression of desmoglein 3 (Dsg3) in mucosal epithelia, which compensates for the loss of Dsg1. However, in rare cases, this protective mechanism can fail, leading to mucosal involvement.
Here, we report an unusual case of pemphigus foliaceus with persistent oral erosions, illustrating how the clinical spectrum of this condition can occasionally extend beyond its expected boundaries.

Case Report
A 62-year-old woman with well-controlled type 2 diabetes presented with painful, recurrent erosions of the oral mucosa, evolving in flares and remissions for over a year. On examination, the lesions were located on the inner aspect of the lips and gingiva, appearing as erythematous erosions with whitish epithelial collars at the edges. The patient reported significant discomfort, particularly during eating and speaking.
Several months after the initial oral symptoms, she developed post-bullous erosive lesions on the trunk and abdomen. These skin lesions appeared gradually and coalesced into large, superficial areas of denudation.
Biopsies of both skin and mucosal tissue were performed. Histology revealed superficial intraepidermal cleavage with acantholytic keratinocytes. Direct immunofluorescence showed intercellular IgG and C3 deposits in a characteristic “net-like” pattern along the epidermis.
Serologic testing using ELISA identified circulating autoantibodies against Dsg1 (index: 1.5), with no detectable anti-Dsg3 antibodies, confirming the diagnosis of pemphigus foliaceus.
The patient was started on oral corticosteroids at 1 mg/kg/day and promptly received two rituximab infusions (1 g on days 1 and 15), followed by maintenance doses of 500 mg every

six months. Cutaneous lesions improved rapidly, allowing gradual tapering and discontinuation of corticosteroids over six months.

However, three months later, the patient experienced a relapse limited to the oral mucosa. Colchicine was introduced at a dose of 1 mg/day for three months, resulting in a clear and sustained improvement.

Discussion

Mucosal involvement in pemphigus foliaceus is rare but biologically plausible. The classic "compensation model" suggests that the presence of Dsg3 in mucosal tissue protects against blistering when only Dsg1 is targeted. However, this balance can be disrupted under certain conditions — for instance, when autoantibody levels are particularly high or when chronic inflammation alters epithelial permeability.

Recent studies have shown that even in cases restricted to anti-Dsg1 autoimmunity, mucosal epitopes may become accessible to circulating antibodies, especially during prolonged disease activity. Direct immunofluorescence has occasionally revealed IgG deposits on mucosal biopsies in pemphigus foliaceus, supporting the idea that the mucosal barrier is not always impenetrable to Dsg1-directed autoimmunity.

This case underscores the need to consider pemphigus foliaceus in the differential diagnosis of chronic oral erosions, even when mucosal involvement seems atypical. It also highlights the effectiveness of rituximab in managing both cutaneous and mucosal disease, and suggests a potential role for colchicine as an adjunct therapy in isolated relapses.

Conclusion

Although rare, mucosal involvement can occur in pemphigus foliaceus when the compensatory role of desmoglein 3 fails to contain the pathogenic effects of anti-Dsg1 autoantibodies. This case illustrates an uncommon but clinically important variant of the disease. A tailored treatment approach — combining systemic immunosuppression and adjunctive therapies like colchicine — allowed successful control of both the initial presentation and subsequent mucosal relapse.

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