A new emergence of pemphigus vulgaris

Pemphigus is an autoimmune disease with intraepidermal bullous lesions, which may affect the oral, genital, or ocular mucosa and the skin. It usually affects people between the ages of 50 and 60 years and is generally evenly distributed between the sexes. The triggering mechanism that initiates the immune response is not known yet.\textsuperscript{1,2} Pemphigus vulgaris is the most common form of pemphigus. The lesions are often insidious and localized. Persistent, painful ulcers and a burning sensation, which affect the appetite, are the clinical findings in pemphigus vulgaris. Mucosal and skin surfaces may be involved in severe cases. Moreover, the conjunctival, pharyngeal, and laryngeal mucosa may be involved, along with extensive skin lesions. The diagnosis of pemphigus vulgaris is made from mucosal biopsies and subsequent histological examination.\textsuperscript{3,4} Furthermore, fluorescent techniques are used for the detection and localization of the tissue-bound antibodies. Nikolsky’s sign is highly indicative of pemphigus that might be absent in some patients with pemphigus vulgaris.\textsuperscript{4} The process of destruction of the intercellular connections (desmosomes) of the epithelial cells (acantholysis) has been observed in these cases. Antibodies against desmoglein 3, generally IgG, can be found tissue-bound, at the level of the spinous cell membrane and circulating in the blood of patients with active pemphigus vulgaris.\textsuperscript{4,5}

This case report describes a new emergence of pemphigus vulgaris, with an erosive, bullous and ulcerative form involving mucosal surfaces. It appeared that this case might be a relatively new case of pemphigus vulgaris. A 60-year-old man came to the Oral Medicine Department of Shiraz School of Dental Medicine, with the complaint of

![Figure 1](A) Oral mucosa and lower lip clinical photographs of the patient at initial presentation. (B) Histopathological examination of the specimen showing suprabasal split and acantholysis. (C) Significant healing of oral mucosal ulcerations after 3 weeks of treatment.

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bleeding ulcers, together with a moderate level of pain and burning sensation in the oral mucosa. The patient explained that a blister formed on his lip about 7 months previously after tooth extraction, which then changed to ulcers on the lips that remained without improvement. He mentioned that few dentists and physicians had seen him. However, none of the procedures and medications was able to improve the situation. Upon clinical examination, there were ulcers on the buccal mucosa, ventrolateral aspect of the tongue, and upper labial mucosa, as well as bleeding ulcers and crusts on the lower lip with a keratinized zone at all these regions, especially the buccal mucosa (Fig. 1A). The patient was negative for Nikolsky’s sign. Subsequently, a biopsy was taken from the ulcerative and keratinized area of the right buccal mucosa and the histopathological examination confirmed the diagnosis of pemphigus vulgaris (Fig. 1B). Additionally, an immunofluorescence study supported the histopathological diagnosis, showing positive IgG antibody binding on the cell membrane of spinous cells. The patient was treated with 50 mg prednisolone, 50 mg azathioprine, and intravenous triamcinolone. After 1 week of treatment, significant improvement was observed. At 6 months follow-up, the oral mucosa showed significant healing and the patient did not report any pain or discomfort (Fig. 1C).

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References


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