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Mucosal-dominant pemphigus vulgaris in a 90-year-old woman

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DESCRIPTION

A 90-year-old Japanese woman was referred to our department with a 3-month history of painful gingivae that first appeared at age 90 years. She reported difficulty in eating because of multiple painful erosions following easily ruptured vesicles in the buccal mucosa and tongue during the last 3 weeks. Her medical history included hyperlipidaemia and reflux esophagitis, for which she regularly consumed atorvastatin calcium hydrate and esomeprazole.

A general examination revealed a few erythematous erosions on the back. An intraoral examination revealed multiple erosions and desquamation with erythematous areas on the right and left buccal mucosa, palate, lateral border of the tongue

and gingiva (figure 1A–C). She was positive for Nikolsky's sign. Differential diagnosis included oral lichen planus, mucous membrane pemphigoid and pemphigus vulgaris (PV). A gingival biopsy for histopathology revealed intraepithelial cleavage with acantholysis in the suprabasal region and retaining of basal keratinocytes along the basement membrane zone (figure 1D). Direct immunofluorescence study showed the deposition of IgG and C3 between the epithelial cells (figure 1E,F). The indirect immunofluorescence ELISA for desmoglein-3 and desmoglein-1 antibodies revealed their elevated levels (126 U/mL and 21 U/mL, respectively, reference range for both is <20). Based on all clinical, histopathological and serological findings, we confirmed mucosal-dominant PV with minimal skin lesions.

The patient received oral prednisolone at an initial dosage of 35 mg/day (1 mg/kg/day). This helped in rapid epithelialisation of the erosions on the back. The intraoral erosions diminished significantly in 2 weeks, and the dose was tapered to 30 mg/day. A marked improvement in the lesions was observed on subsequent follow-up. The patient is currently on prednisolone treatment (15 mg/day), and to date, no other lesions have been reported.

PV is an autoimmune disorder of the skin and mucous membranes that triggers intraepithelial blistering and usually develops in adults aged 40–60 years.¹ Ojaimi *et al* have reported two cases of mucosal-predominant PV newly diagnosed at age 70 and 75 years.² PV is scarcely reported in the elderly, especially in the oldest-old (those aged ≥85 years) patients.^{3,4} To the best of our knowledge, this patient would be the oldest individual with mucosal-dominant PV reported to date in English literature. The oral mucosa is frequently affected. Mucosal lesions are the first manifestations of PV in most patients. In some patients, however, the cutaneous lesions follow oral manifestations. Hence,

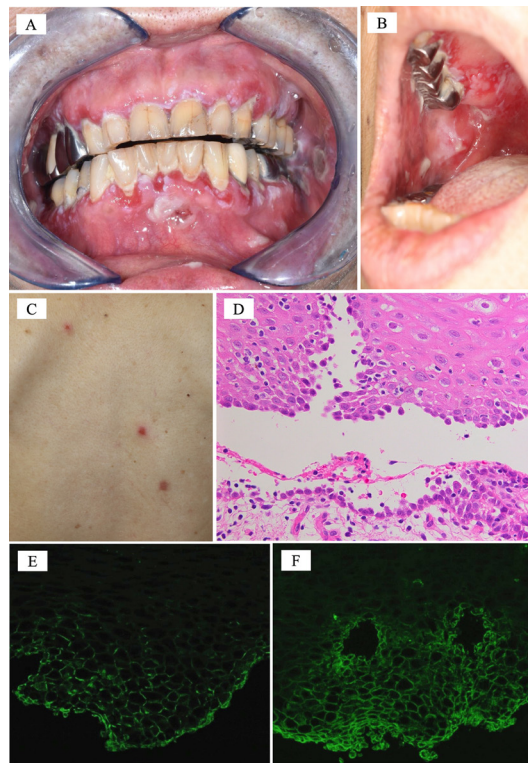


Figure 1 (A) Swollen gingivae surrounding the upper and lower front tooth with an erosion on the lower gingiva. (B) Erosions seen on the buccal mucosa. (C) Minimal erosions affecting the cutaneous surface of the back. (D) Histopathology of the buccal mucosa revealing intraepithelial cleavage with suprabasal acantholysis (H&E staining; original magnification: $\times 400$). (E) Low power view ($\times 20$) of direct immunofluorescence staining for IgG. (F) Low power view ($\times 20$) of direct immunofluorescence staining for C3.

Learning points

- ▶ Pemphigus vulgaris (PV) rarely affects the oldest-old population.
- ▶ Refractory oral membrane erosions are the presenting signs of PV and should raise suspicion of this disease.
- ▶ Early diagnosis and initiation of treatment with systemic glucocorticoids, the mainstay of therapy for PV, is usually highly effective for controlling the disease.



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timely diagnosis and treatment of oral lesions can avert skin involvement.

Early diagnosis is challenging since the oral lesions may be relatively non-specific, emerging as superficial erosions or ulcerations and rarely manifesting as intact bullae.⁵ Although the lesions could be anywhere on the oral mucosa, they are commonly found over the buccal mucosa, palate, tongue and lip, and less frequently at the gingiva.⁶

Physicians should be sufficiently familiar with the oral manifestations of PV, such as gingivitis, even in the oldest older patients, and should involve specialists to ensure early diagnosis and treatment, thereby influencing the prognosis and course of this disorder.

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Contributors TM conducted the review of the patient's case notes, review of the literature and the writing of the clinical image. SK, YS and MK was responsible for identifying and managing the case, as well as reading and suggesting improvements to several drafts, and in agreeing on the final report.

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