

## CORRESPONDENCE

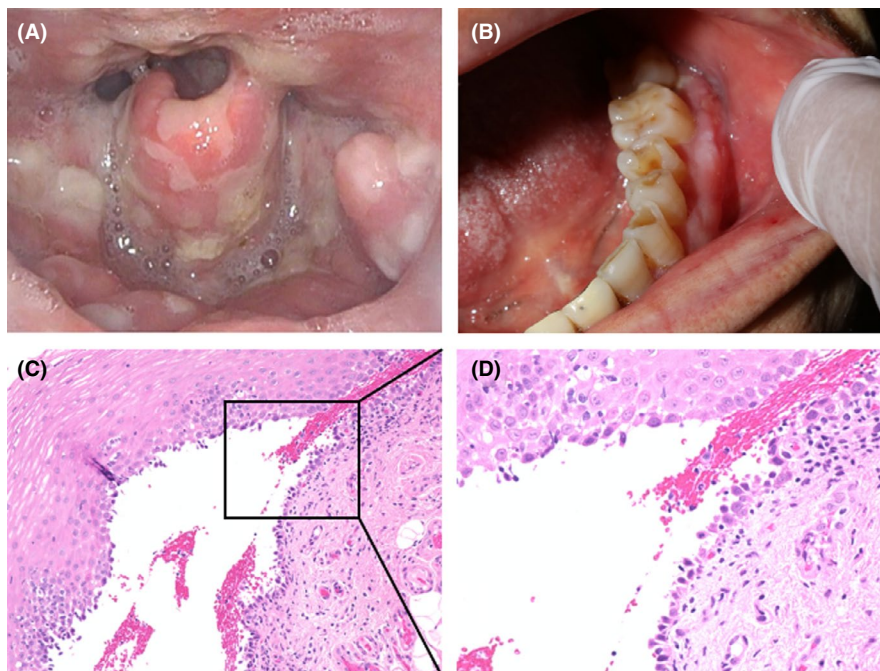
## Primary laryngeal pemphigus vulgaris with severe epiglottitis

Dear Editor,

An 80-year-old man visited a private otolaryngology clinic for sore throat and was treated with oral antibiotics for 1 week without any improvement. The lack of response to treatment prompted the referral to the department of otorhinolaryngology at our hospital. At the first visit, the patient presented with hoarseness, hemoptysis, and dysphagia. The oral cavity and skin were unaffected. Laryngoscopy revealed severe supraglottic inflammation with mucosal erosion covering almost the entire area of the oropharynx and larynx. His upper airway was constricted due to prominent edema of the epiglottis (Figure 1A). These findings were consistent with a diagnosis of acute epiglottitis. Treatment with intensive intravenous corticosteroids and antibiotics resulted in temporary improvement of the inflammation. However, multiple

mucosal erosions appeared on the lip, soft palate, and gingiva upon tapering the dose of intravenous corticosteroids (Figure 1B). As the oral cavity is rarely impaired in acute epiglottitis and the mucosal erosions were resistant to the initial treatment, other disorders were suspected.

A biopsy of the epiglottis was performed, and histopathological findings revealed acantholysis of the mucosal epithelium and suprabasal blister formation (Figure 1C,D). There was no increase in anti-desmoglein-1 antibody level. However, the anti-desmoglein-3 antibody level was elevated (43.4 U/ml). These histological and serological findings were consistent with a diagnosis of pemphigus vulgaris (PV) with severe epiglottitis. Treatment with oral prednisolone (60 mg, daily) gradually improved the symptoms. Follow-up laryngoscopy revealed the resolution of the supraglottic inflammation



**FIGURE 1** Clinical and histopathological findings of the patient. Laryngoscopy revealed erythematous supraglottic edema with multiple mucosal erosions covered by whitish exudates seen in almost all of the oropharynx and larynx. The epiglottis showed prominent edema (A). Erosions on the left gingiva appeared during the initial treatment (B). A biopsy of the epiglottis revealed suprabasal blister formation demonstrating a “row of tombstones appearance” and acantholysis of the suprabasal layer cells (hematoxylin and eosin stain, original magnification, C  $\times 100$ /D  $\times 200$ )

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and erosions. Systemic steroids were gradually reduced, and the PV was well controlled for more than 6 months.

Pemphigus vulgaris is an autoimmune bullous disease characterized by intraepithelial blistering of the skin and mucous membranes caused by autoantibodies against desmosomal components.<sup>1</sup> To date, primary laryngeal PV has rarely been reported, and although delayed diagnosis is common even in primary oral PV, primary laryngeal PV without oral or skin lesions is believed to be more challenging to diagnose.<sup>2-4</sup> It may be misdiagnosed as bacterial or viral laryngitis, such as acute epiglottitis, due to its rarity, and inappropriate treatment may be administered. In our case, intensive intravenous corticosteroids were administered at a dose appropriate for the treatment of acute epiglottitis, and the dose was reduced more rapidly than suitable for common pemphigus, which may have caused the symptoms to flare up. However, a failure to administer corticosteroids would have worsened the epiglottic edema, causing upper airway obstruction. It should be noted that preserving the upper airway in patients with PV is more difficult because of the Nikolsky phenomenon. The pharyngeal and laryngeal mucosa are fragile and may easily be impaired by mechanical irritation of the endotracheal tube.<sup>5</sup> This means that severe epiglottitis in patients with PV can be lethal. For primary laryngeal PV, dermatologists and otorhinolaryngologists should work closely to properly evaluate and manage the oral cavity, larynx, and pharynx, followed by early diagnosis and initiation of urgent, timely, and appropriate treatment to reduce the risk of life-threatening upper airway obstruction.

#### DECLARATION SECTION

Approval of the research protocol: N/A.


Informed Consent: Written informed consent was obtained from the patient.


Registry and the Registration No.: N/A.

Animal Studies: N/A.

#### CONFLICT OF INTEREST

The authors declare no conflict of interest.

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