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Pemphigus Vulgaris Presenting with Initial Dermatological Manifestations and Occult Laryngeal Involvement: A Rare ENT-Dermatology Correlated Case

Dr. Ankush Babal[1], Dr. Pooja Saroj[2]

[1]Department of Dermatology, Venereology and Leprosy, F H Medical College & Hospital, Agra, India
Email: *ankushbabal01234[at]gmail.com*

[2]Department of Otorhinolaryngology, F H Medical College & Hospital, Agra, India
Email: *pooja.saroj0208[at]gmail.com*

Abstract: ***Pemphigus vulgaris is a rare, potentially life-threatening autoimmune blistering disorder predominantly affecting the skin and oral mucosa. Laryngeal involvement is uncommon and often underdiagnosed due to subtle or nonspecific symptoms. We report a case of a 38-year-old female who presented with classical cutaneous and oral manifestations of pemphigus vulgaris and was incidentally found to have occult laryngeal involvement on otorhinolaryngological evaluation. Dermatological investigations including Tzanck smear, histopathology, and direct immunofluorescence confirmed the diagnosis. Flexible fiberoptic laryngoscopy revealed superficial erosions over the supraglottic structures without airway compromise. Early initiation of systemic corticosteroids and rituximab led to rapid clinical remission of both cutaneous and laryngeal lesions. This case highlights the importance of a high index of suspicion and routine ENT evaluation in patients with pemphigus vulgaris, even in the presence of minimal laryngeal symptoms, to prevent potentially life-threatening airway complications. Close interdisciplinary collaboration ensures early diagnosis and favorable outcomes.***

Keywords: Pemphigus vulgaris; Laryngeal involvement; Autoimmune blistering disease; Hoarseness of voice; ENT–Dermatology interface; Occult airway involvement

1. Introduction

Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder characterized by circulating autoantibodies directed against desmoglein 3 and desmoglein 1, resulting in suprabasal acantholysis. Clinically, PV commonly presents with mucocutaneous involvement, with painful oral erosions often preceding cutaneous lesions. Although the oral cavity is the most frequently involved mucosal site, extension to the pharynx and larynx is rare and usually associated with advanced disease.

Laryngeal involvement may remain subclinical or present with mild symptoms such as hoarseness, leading to delayed diagnosis. Early recognition is essential, as progressive laryngeal disease can result in airway compromise. This case report emphasizes the importance of interdisciplinary evaluation by dermatology and otorhinolaryngology services in detecting occult laryngeal involvement in PV.

2. Case Report

A 38-year-old female presented to the Dermatology outpatient department with a three-week history of painful oral erosions and multiple fluid-filled lesions over the trunk, associated with burning sensation and difficulty in eating. The blisters were flaccid, ruptured easily, and healed with raw erosions. There was no history of fever, recent drug intake, or similar illness in the past. The patient also reported mild hoarseness of voice for a few days, which she initially ignored.

3. Dermatological Examination

Cutaneous examination revealed multiple flaccid bullae and erosions over the chest, back, and upper limbs, with irregular margins and surrounding erythema. Nikolsky's sign was positive. Oral examination showed extensive erosions involving the buccal mucosa, gingiva, and soft palate. Based on these findings, a provisional diagnosis of pemphigus vulgaris was made.

4. Dermatological Investigations

Tzanck smear from a fresh blister demonstrated numerous acantholytic cells. Histopathological examination of a perilesional skin biopsy revealed supranasal cleft formation with acantholysis, producing a characteristic "row of tombstones" appearance. Direct immunofluorescence showed intercellular deposition of IgG and C3 in a fish-net pattern, confirming the diagnosis of pemphigus vulgaris.

5. ENT Evaluation

In view of the history of hoarseness, the patient was referred for ENT assessment. Indirect laryngoscopy showed mild erythema and edema of the supraglottic mucosa with normal vocal cord mobility. Flexible fiberoptic laryngoscopy revealed superficial erosions over the epiglottis and arytenoids with mild mucosal sloughing. The lesions were friable but did not cause airway narrowing. These subtle findings would likely have been missed without targeted examination.

6. Treatment

Systemic corticosteroid therapy was initiated with oral prednisolone at 1 mg/kg/day. Considering the presence of laryngeal involvement and the risk of airway compromise, early biologic therapy with rituximab was started following the rheumatoid arthritis protocol (1 g intravenously on day 1 and day 15). High-potency topical corticosteroids were prescribed for cutaneous lesions, and topical anesthetic gels were used for oral erosions. Supportive measures included calcium and vitamin D supplementation and proton pump inhibitors.

From the ENT perspective, the patient was advised voice rest and avoidance of airway irritants. Serial flexible laryngoscopic examinations were performed to monitor lesion resolution, and the patient was counseled regarding warning symptoms of airway compromise.

7. Follow-Up

At two-week follow-up, the patient showed significant improvement with healing of oral and cutaneous lesions and complete resolution of hoarseness. Repeat laryngoscopy demonstrated marked improvement with healing of supraglottic erosions. At one-month, cutaneous lesions had healed with post-inflammatory hyperpigmentation, and systemic steroids were gradually tapered. At six months, the patient remained in complete remission without recurrence of dermatological or laryngeal disease.

8. Discussion

Although pemphigus vulgaris is primarily a dermatological condition, mucosal involvement may extend beyond the oral cavity to involve the larynx. Laryngeal lesions are often underdiagnosed due to mild or nonspecific symptoms. This case underscores the importance of routine ENT evaluation in PV patients, even when laryngeal symptoms are minimal. Early diagnosis and prompt systemic therapy can prevent progression to severe complications such as airway obstruction. Interdisciplinary collaboration is crucial for optimal patient outcomes.

9. Conclusion

Pemphigus vulgaris may present with classic mucocutaneous lesions while laryngeal involvement remains occult. A high index of suspicion and timely ENT evaluation are essential for early detection of airway involvement. Coordinated management between dermatologists and otorhinolaryngologists leads to favorable outcomes and prevents life-threatening complications.

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