Abstract

- **Objectives:** To discuss the manifestations of pemphigus vulgaris in the upper aerodigestive tract and the role of the otolaryngologist.
- **Study Design:** Case report and literature review.
- **Methods:** This is a case report describing a patient with pemphigus vulgaris affecting the larynx and manifesting as signs concerning for epiglottitis.
- **Results:** The diagnosis was made based on history, exam, and biopsy with positive direct immunofluorescence of immunoglobulin in squamous epithelium. Furthermore, the literature review demonstrates that a large proportion of patients may have laryngeal findings, even if they deny throat symptoms.
- **Conclusions:** Therefore, evaluation by an otolaryngologist is essential in the initial evaluation to aid in diagnosis and to monitor the response to treatment.

Introduction

- Pemphigus vulgaris (PV) is a rare bullous disorder of mucosa and skin. PV is caused by antibodies against desmoglein, a glycoprotein integral to intercellular adhesion in epidermis and produces intra-epidermal cleavage in deepest layers of epidermis and cause flask blisters.
- PV manifests in mucosal lesions, cutaneous lesions, or both.
- Dermatologists primarily manage PV with systemic glucocorticoids, with or without immunomodulatory agents.
- Otolaryngology literature review demonstrates evidence of PV affecting upper aerodigestive tract as early as 1899.
- Multiple studies have demonstrated further evidence of PV causing mucosal lesions of nasal cavity, oral cavity, oropharynx, and larynx based on physical exam and laryngoscopy, even in asymptomatic patients.
- The most common location for lesions is in the oral cavity.
- Mahmoud et al has described a classification system to determine the most common sites of lesions in the larynx.
- Most laryngeal lesions were localized to the epiglottis (denoted Grade I).
- Studies demonstrate that medical treatment of PV with steroids and immunosuppressant is effective in improving mucosal lesions at all upper aerodigestive tract sites.
- Literature review recommends otolaryngology evaluation for all patients with PV.

Case Report

- 38 year old man presented to the emergency department with throat pain and odynophagia.
- Otolaryngology was consulted to evaluate for “epiglottitis.”

Results

- The diagnosis of PV was based on history, exam, direct laryngoscopy, and oral pathology findings.
- Physical exam demonstrated mucosal ulceration of buccal and floor of mouth mucosa (Figure 1).
- Flexible laryngoscopy demonstrated ulceration of bilateral arypeiglottic folds, left false vocal fold, lingual epiglottis, and perynoid sinuses (Figure 2).
- Biopsy was performed and results showed evidence of intraepidermal cleavage and “lace-like” pattern of immunoglobulin deposition between epidermal cells on direct immunofluorescence (Figure 3 and Figure 4).
- Patient admitted to hospital for airway observation and management with high dose steroids.
- After discharge patient began rituximab infusions and improved with high dose steroids.
- Additional treatment was required to reevaluate and monitor the response to treatment.

Discussion

- Patients with PV may manifest mucosal lesions, cutaneous lesions, or both.
- The most common location for mucosal lesions is the oral cavity.
- Patients with PV are primarily managed by dermatologists, however, mucosal lesions may also extend to the larynx causing odynophagia, hoarse voice, and even respiratory distress.
- Furthermore, the literature review demonstrates that a large proportion of patients may have laryngeal findings, even if they deny throat symptoms.
- Therefore, evaluation by an otolaryngologist is essential in the initial evaluation to aid in diagnosis and to monitor the response to treatment.

Conclusion

This case demonstrates the importance of a multidisciplinary approach to treating pemphigus vulgaris of the upper aerodigestive tract and highlights the critical role of the otolaryngologist in the management of the disease.

References