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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 immunofluorescent studies. The patient has responded well to standard therapy and has required routine otolaryngology follow-up.
- **Conclusions:** Patients with pemphigus vulgaris may manifest mucosal lesions in the oral cavity and larynx. Evaluation by an otolaryngologist is essential in the initial evaluation and to monitor the response to treatment. This case demonstrates the importance of a multidisciplinary approach to treating pemphigus vulgaris of the upper aerodigestive tract.

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 flaccid 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 1899.¹
- 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.^{2,3}
- The most common location for lesions is in the oral cavity.³
- Mahmoud et. al has described a classification system to determine the most common sites of lesions in the larynx.⁴
- Most laryngeal lesions were localized to the epiglottis (denoted Grade 1).⁴
- Studies demonstrate that medical treatment of PV with steroids and immunosuppressant is effective in improving mucosal lesions at all upper aerodigestive tract sites.⁵
- Literature review recommends otolaryngology evaluation for all patients with PV.²⁻⁵

Case Report

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

Figure 1. Oral Cavity Exam.

- (a) Floor of mouth and ventral tongue ulcerative lesions
- (b) Buccal mucosa ulcerative lesions

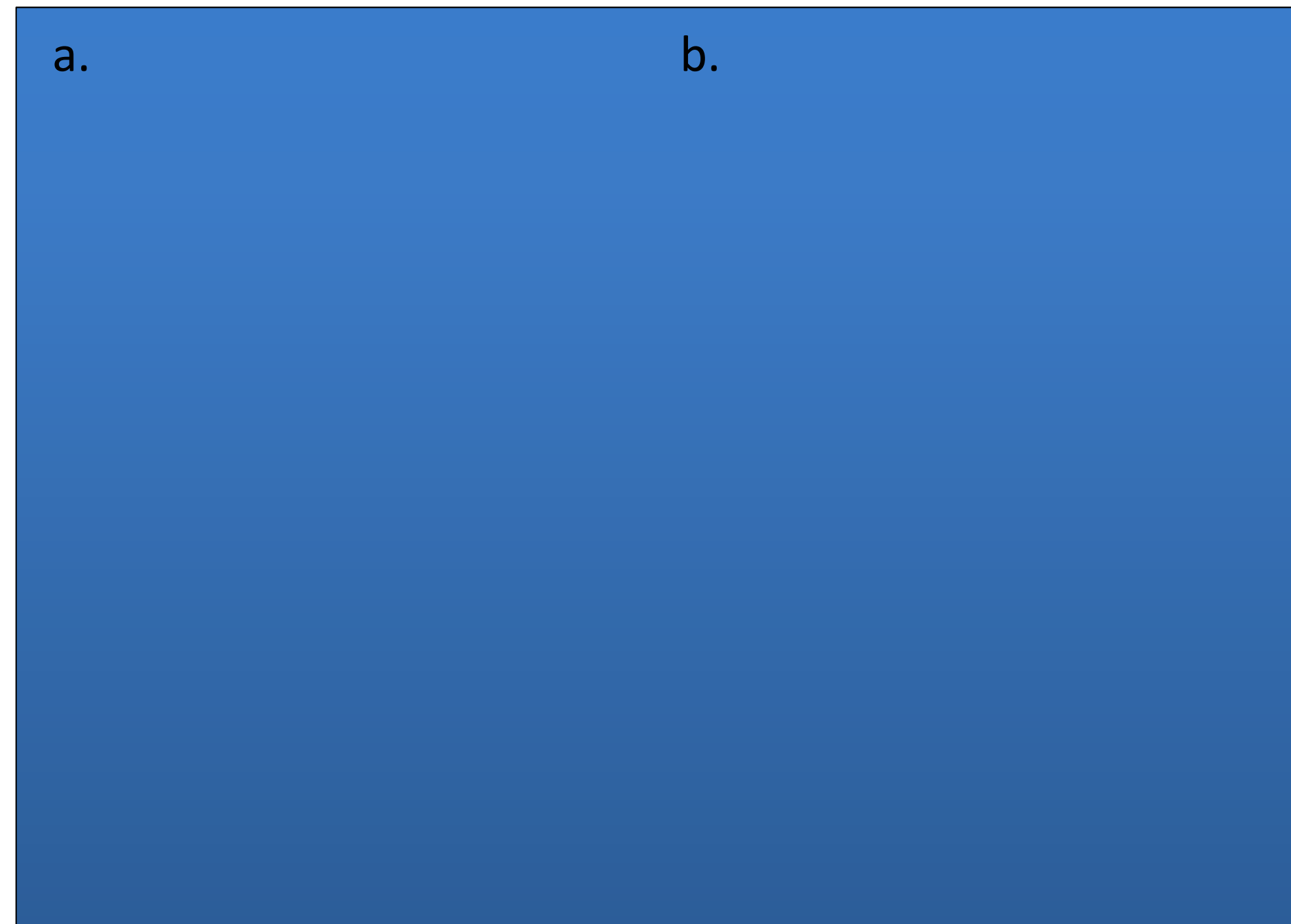


Figure 2. Flexible Laryngoscopy.

- (a) Bilateral aryepiglottic fold ulcerations, Left false vocal fold ulceration
- (b) Thickened epiglottis with ulceration on lingual surface

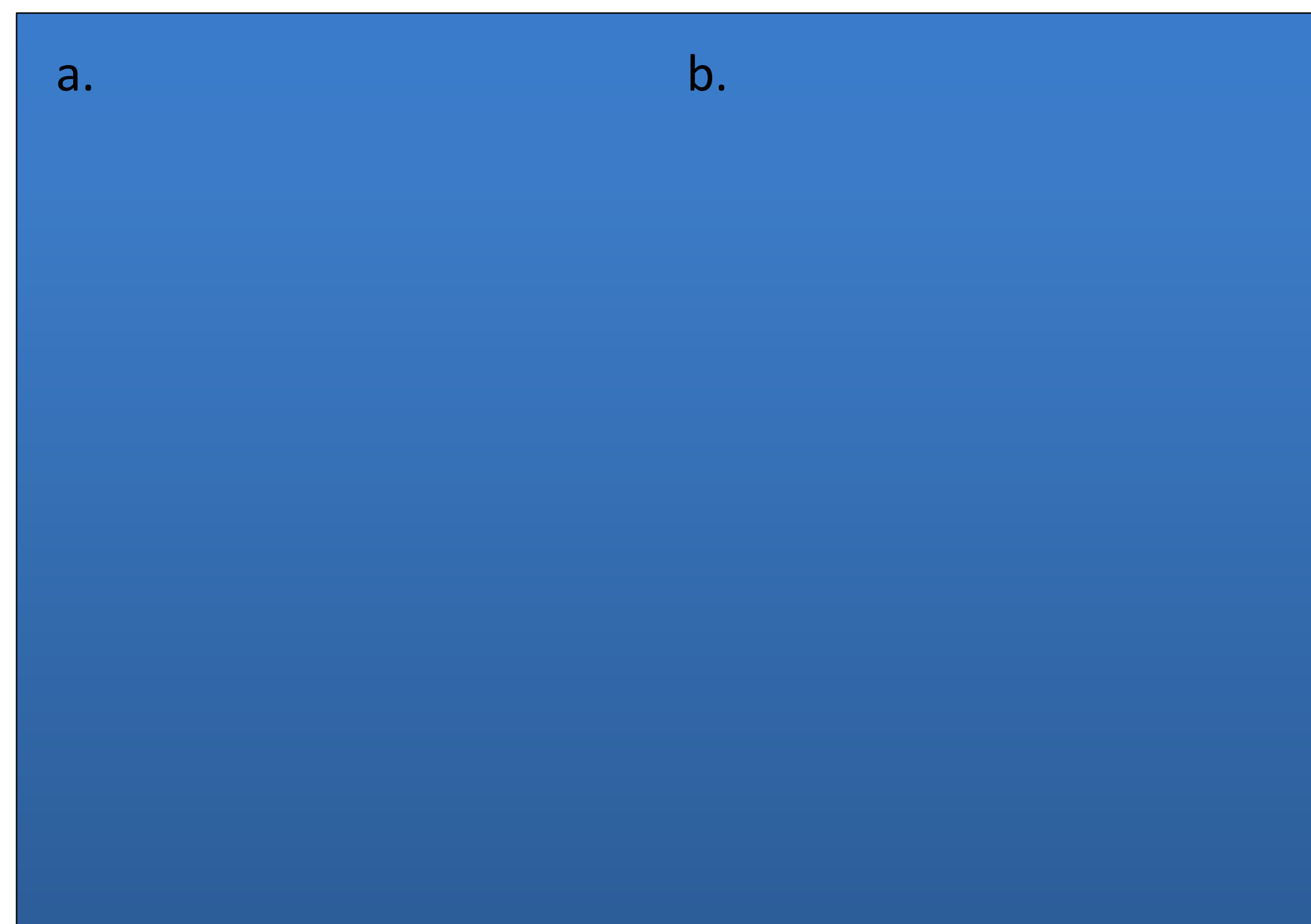


Figure 3. H&E Stain of Oral Mucosal Lesion. Intraepidermal cleavage and loss of the suprabasal strata of the squamous epithelium.

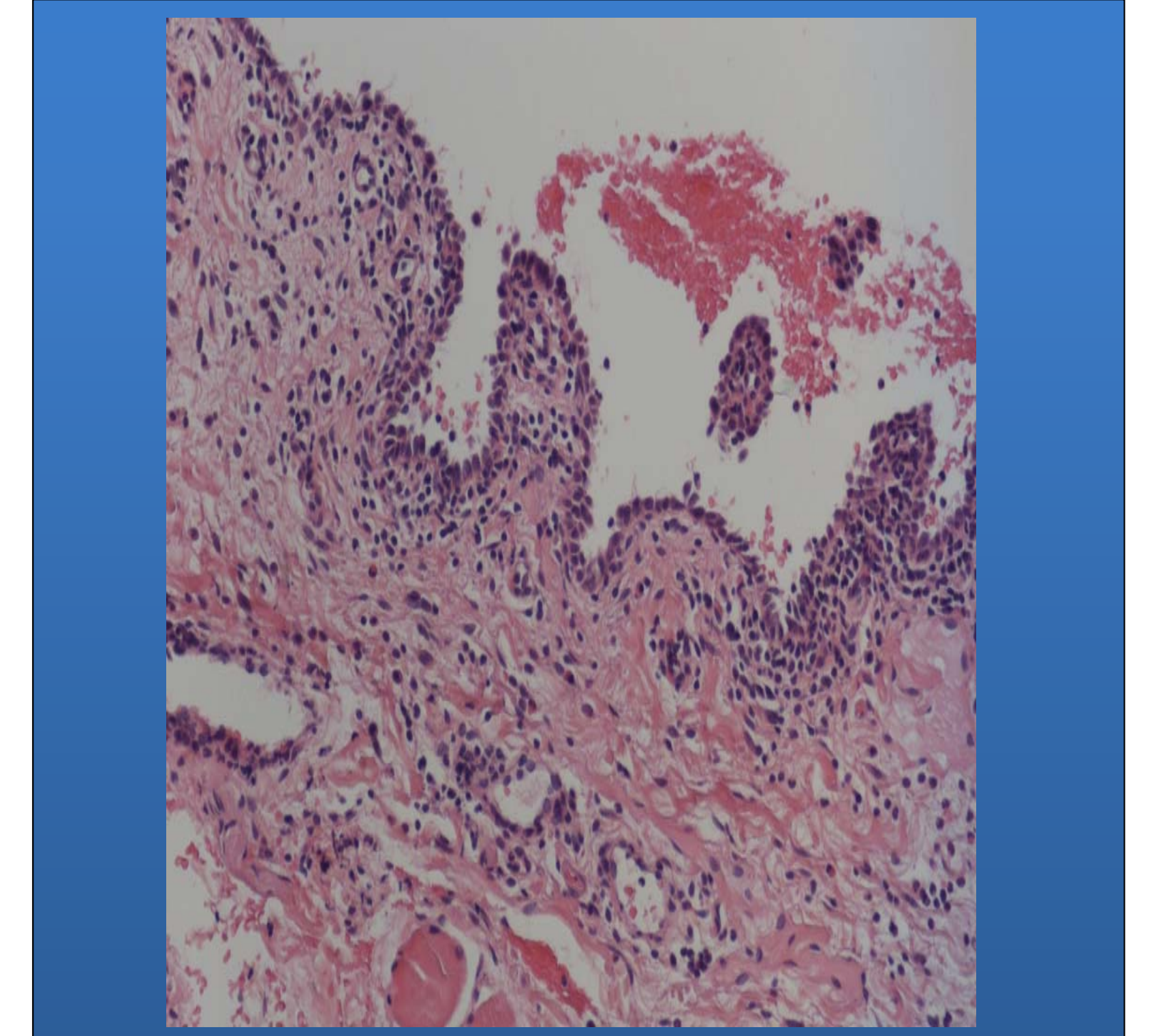
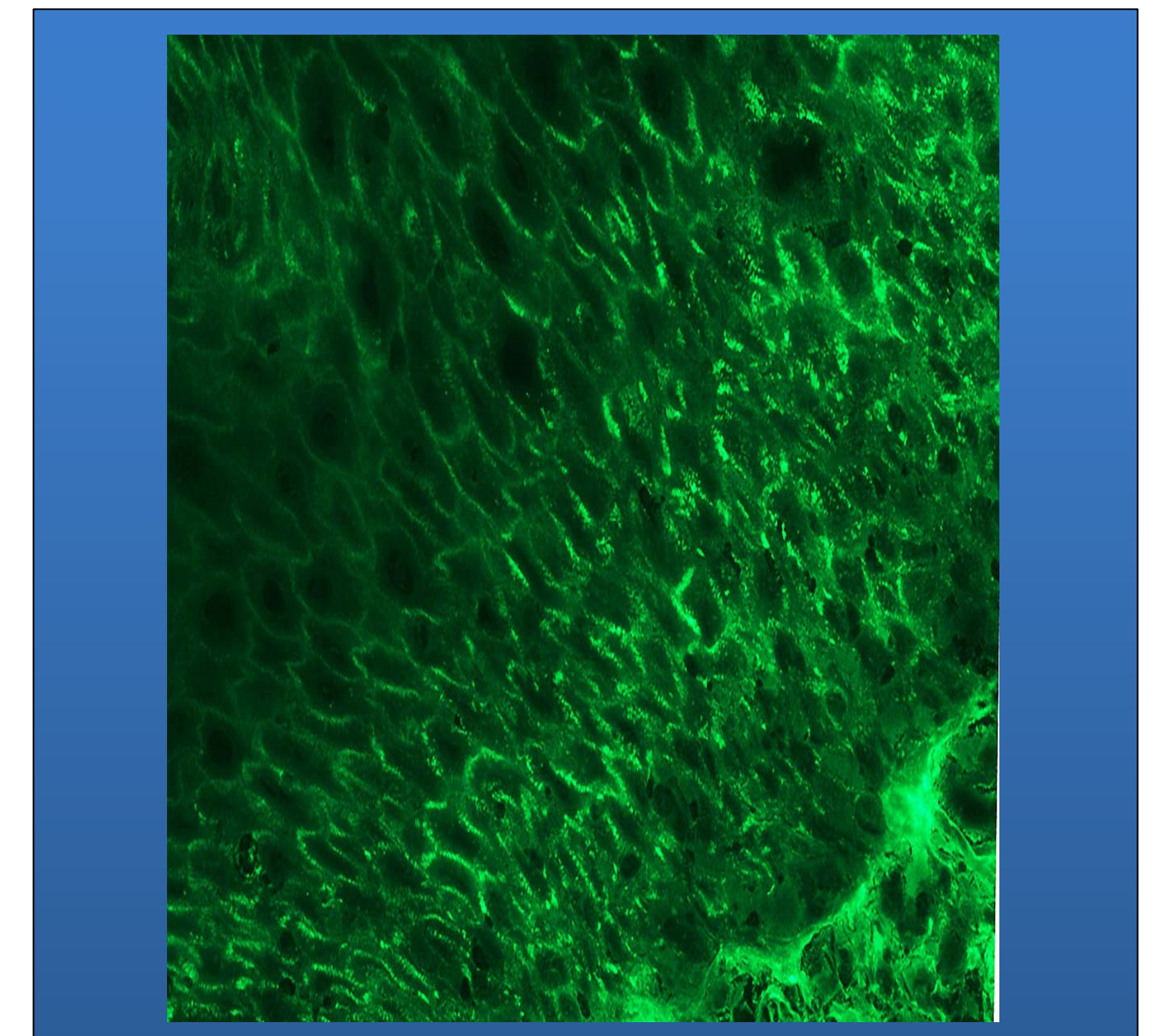


Figure 4. Direct Immunofluorescence. Intercellular fluorescence of immunoglobulin in squamous epithelium.



Results

- The diagnosis of PV was made 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 aryepiglottic folds, left false vocal fold, lingual epiglottis, and pyriform 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 prednisone.
- After one month of treatment, patient was reevaluated and showed significant improvement of symptoms and mucosal lesions.
- He continues to require routine follow up with otolaryngology for recurrent oral cavity, oropharyngeal, and laryngeal lesions in between infusions.

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.^{2,3}
- 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.

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