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CASE REPORT

Isolated Laryngeal Involvement in Mucosal Membrane Pemphigoid: Diagnostic and Surgical Treatment Approach

Mukozal Membran Pemfigoidinde İzole Larenks Tutulumu: Tanısal ve Cerrahi Tedavi Yaklaşımı

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ABSTRACT

Aim: Mucous membrane pemphigoid (MMP) is a chronic autoimmune subepithelial blistering disorder that primarily affects mucosal surfaces. Laryngeal involvement is uncommon but clinically important, as it may result in progressive scarring and severe airway obstruction. Because of its rarity, the diagnosis can be easily overlooked, and the number of reported cases in the literature remains limited. In this case report, we present the clinical findings, diagnostic approach, treatment strategy, and follow-up of a patient with laryngeal MMP. By sharing this case, we aim to contribute to the limited body of evidence on this rare presentation and highlight the importance of early recognition and appropriate management in preventing life-threatening complications.

Keywords: Pemphigoid; Autoimmune Diseases; **Laryngeal Mucosa;** Laser.

ÖZ

Amaç: Mukoza zarı pemfigoidi (MMP), öncelikli olarak mukozal yüzeyleri etkileyen kronik bir otoimmün subepitelyal büllöz hastalıktır. Larenks tutulumu nadirdir, ancak klinik açıdan önemlidir, çünkü ilerleyici skarlaşma ve ciddi hava yolu tıkanıklığına neden olabilir. Nadir görülmesi nedeniyle tanı kolayca gözden kaçabilir ve literatürde bildirilen vaka sayısı sınırlıdır. Bu olgu sunumunda, larengeal MMP'li bir hastanın klinik bulgularını, tanı yaklaşımını, tedavi stratejisini ve takibini sunuyoruz. Bu vakayı paylaşarak, bu nadir tabloya ilişkin sınırlı sayıdaki kanıta katkıda bulunmayı ve yaşamı tehdit eden komplikasyonları önlemede erken tanı ve uygun tedavinin önemini vurgulamayı amaçlıyoruz.

Anahtar Kelimeler: Pemfigoid; Otoimmün Hastalıklar, Laryngeal Mucosa; Laser

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INTRODUCTION

Mucous Membrane Pemphigoid (MMP) is a rare, chronic autoimmune blistering disorder classified within the pemphigoid group of diseases (1).The disease most commonly involves the ocular and oral mucosa, where it presents with bullae (2). Patients may present with mild symptoms such as gingival erythema and oral lesions, but in some cases, severe tracheal stenosis leading to respiratory distress can also be the clinical symptom (1). The incidence of laryngeal involvement in mucous membrane pemphigoid (MMP) is approximately 12.2%, with the supraglottic region being the most commonly affected site, observed in 84.4% of cases (1). The aim of this case report is to contribute to the existing literature by presenting a rare case of laryngeal mucous membrane pemphigoid (MMP).

CASE REPORT

A 74-year-old female patient presented to our clinic with complaints of respiratory distress and dysphagia. The patient's medical history revealed that her symptoms had been ongoing for the past 1–2 years. It was noted that she experienced recurrent oral ulcerations and had received pharmacological treatment for this condition. The patient was diagnosed with cicatricial pemphigoid approximately five years ago by the dermatology department at another institution. At the time of presentation, her current medications included azathioprine (50 mg twice daily) and prednisolone (8 mg once daily).

The patient exhibited inspiratory stridor that was present at rest and exacerbated with exertion. No lesions were observed in the oropharyngeal region. Flexible fiberoptic laryngoscopy revealed posterior displacement of the epiglottis, circumferential fibrotic stenosis at the laryngeal inlet involving the arytenoid and epiglottic regions, epithelial thickening of the epiglottic and laryngeal mucosa, and pooling of saliva in the piriform sinuses. The rima glottidis was narrowed to approximately 5 × 5 mm (Figure 1). According to previous medical records, a biopsy of the same lesion had been performed at an external center, which demonstrated infiltration of polymorphonuclear leukocytes (PMNLs). Contrast-enhanced neck computed tomography (CT) revealed no contrast-enhancing mass within the laryngeal structures. Dermatological examination showed no lesions on the skin or other mucosal surfaces. Based on the patient's current clinical findings, systemic corticosteroid therapy was initiated at a dose of 1 mg/kg administered parenterally.

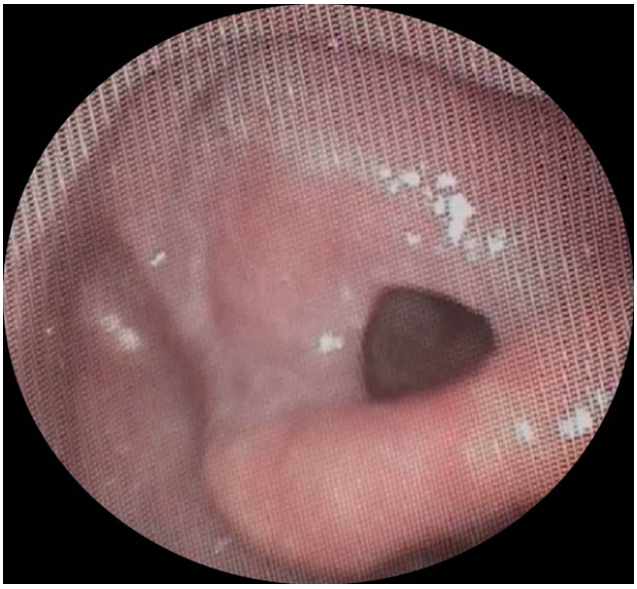

Figure 1: Flexible fiberoptic laryngoscopy revealed circumferential fibrotic stenosis at the laryngeal inlet involving the arytenoid and epiglottic regions, epithelial thickening of the epiglottic and laryngeal mucosa.

During the examination performed under general anesthesia, it was observed that the mucosa separated spontaneously from the

underlying tissue when touched (Nikolsky sign) (Figure 2). A 30-degree endoscopic evaluation confirmed that both vocal cords and the subglottic region were intact (Figure 3). Using a 9-watt continuous-mode CO_2 laser, adhesions between the aryepiglottic folds and the epiglottis were released.

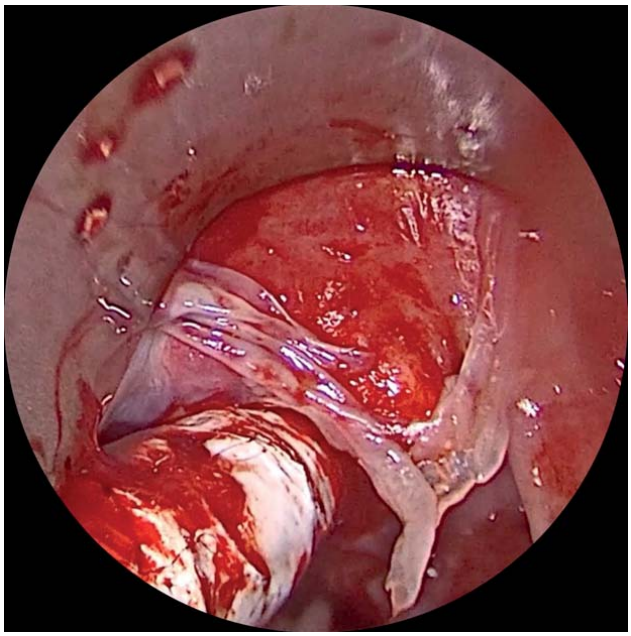

Figure 2: The mucosa spontaneously separated from the underlying tissue when touched (Nikolsky sign).

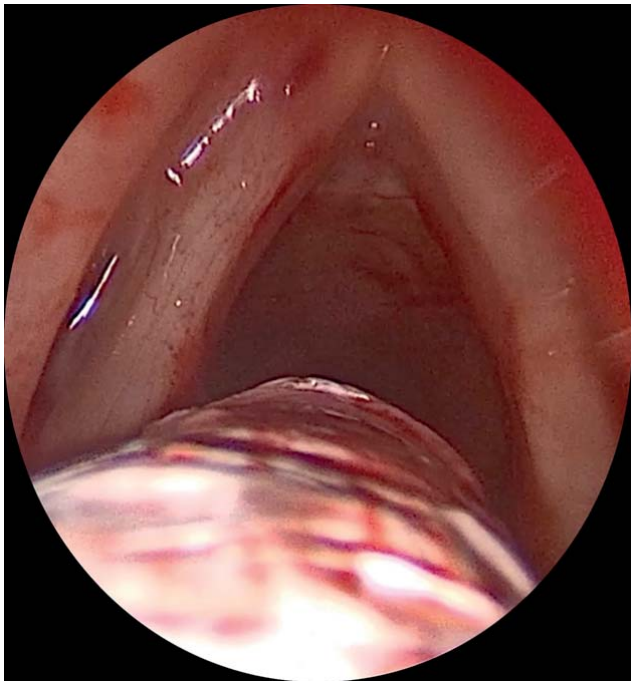

Figure 3: Vocal cords and the subglottic region are observed to be completely normal.

Direct immunofluorescence (DIF) analysis revealed no immunoreactive deposits of IgG, IgA, IgM, or C3. Histopathological examination showed scattered epithelial cell fragments, epithelial hyperplasia, keratosis, and a prominent intraepithelial inflammatory infiltrate rich in leukocytes. Immunohistochemical analysis demonstrated positive staining in the basal layer for Ki-67 and p53.

During postoperative follow-up, the patient attended multiple monthly check-ups and remained free of respiratory distress (Figure 4). Although she did not experience significant difficulties with feeding, aspiration occurred during a meal six months after the operation, resulting in her death at home.

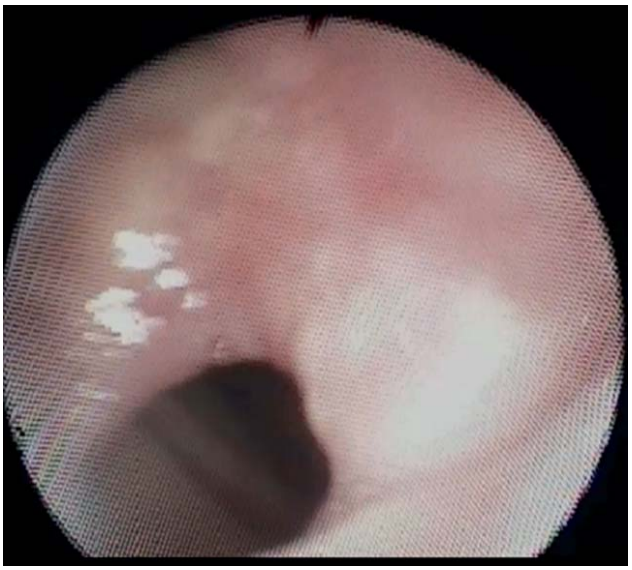

Figure 4: Postoperatively, the laryngeal inlet appears to be significantly relieved.

DISCUSSION

Mucous membrane pemphigoid (MMP) is a rare, chronic autoimmune blistering disorder primarily affecting mucosal surfaces and characterized histologically by subepithelial blister formation. The disease is mediated by autoantibodies directed against various

components of the basement membrane (3). Diagnostic approaches for MMP typically involve a combination of clinical assessment, histopathological analysis, and immunopathological techniques such as direct immunofluorescence (DIF) or direct immunoelectron microscopy (DIEM) (4). According to the MMP consensus recommendations, in instances where DIF from an initial oral mucosal punch biopsy yields negative results, repeat biopsies are strongly recommended to increase diagnostic sensitivity (1). **The sensitivity of DIF varies widely, ranging from 41% to 100%, depending on the biopsy site (5).** There are also studies reporting that immune deposits cannot be demonstrated in patients with laryngeal mucosal involvement (6). In our case, intraoperative findings, such as spontaneous separation of the mucosa from the underlying tissue following mucosal contact, along with histopathological signs of inflammation, and the patient's prior diagnosis of MMP, supported the diagnosis of laryngeal MMP. Consistent with previous reports, direct immunofluorescence (DIF) analysis failed to reveal any immune deposits in the affected tissue. A second biopsy was not performed in our patient due to the established diagnosis and the resolution of respiratory distress following surgical intervention.

PubMed literature review revealed only three reported cases of isolated laryngeal involvement, all of which were classified under the broader category of pemphigoid diseases (7-9). A multidisciplinary approach is essential in the management of mucous membrane pemphigoid (MMP) sym(3). Currently, there are no standardized treatment protocols for laryngeal involvement in mucous membrane pemphigoid (MMP), and existing evidence is primarily derived from isolated case reports. Various therapeutic approaches have been proposed, including corticosteroids, dapsone, methotrexate, plasmapheresis, rituximab, cyclosporine, and intravenous immunoglobulin. In cases complicated by laryngeal stenosis, surgical interventions such as endoscopic laser ablation, airway dilation, or tracheostomy may be necessary to maintain airway patency (10). Consistent with previously reported treatment strategies, our patient underwent perioperative systemic corticosteroid therapy combined with endoscopic laser intervention to achieve adequate supraglottic airway patency.

CONCLUSION

Isolated laryngeal involvement is exceedingly rare, and current literature on its diagnosis, treatment, and long-term follow-up remains poorly documented. This case is important because it emphasizes the need to consider local laryngeal involvement of existing systemic disease in patients with unexplained supraglottic stenosis and a history of autoimmune bullous disease. While respiratory distress often dominates the clinical picture in patients with isolated laryngeal MMP, aspiration and other swallowing-related complications must also be carefully evaluated and monitored throughout the treatment course and follow-up period.

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