Laryngeal Mucous Membrane Pemphigoid (MMP): A Systematic Review and Pooled-Data Analysis.

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ABSTRACT

Objectives: To perform a systematic pooled-data analysis of literature data of laryngeal mucous membrane pemphigoid (MMP).

Study Design: A systematic review and pooled-data analysis.

Methods: We conducted a systematic literature search of MEDLINE, EMBASE, CENTRAL, Cochrane, clinicaltrials.gov, and the National Guideline Clearinghouse databases without language restriction for studies including combinations of relevant terms. All authors independently screened the abstracts of the search results, identified articles eligible for review, and critically appraised the full-text studies. Pooled-data analyses and Kaplan-Meier survival analyses were conducted using SPSS.

Results: Of the 2524 citations reviewed, the included articles consisted of 64 case reports and 10 case series reporting on 141 patients with laryngeal MMP. No clinical trials or comparative trials were found. The overall estimated prevalence of laryngeal MMP was 12.2% (95% CI, 11.5 to 12.9%) of cases of MMP or one in ten-million persons in the general population. The supraglottis was the most commonly affected site (94.8%, 95% CI, 92.5 to 97.2%). Distribution among genders was equivalent (P=0.655). The presence of anti-epiligrin autoantibodies was associated with increased laryngeal involvement (OR 7.9, 95% CI, 3.9 to 16.0). The overall 5-year relative survival rate was 92.4% (SE, 8.4) with a follow-up range of 1 to 221 months. Standard medical therapy alone occasionally improved the condition; however, relapses were frequent and 10.5% eventually required tracheostomy. Laryngeal surgical interventions seemed to be effective in severe cases. Conclusion: Laryngeal MMP is a rare condition that can be life-threatening without proper treatment and frequent follow-up.

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INTRODUCTION

Mucous membrane pemphigoid (MMP) is a relatively rare, chronic autoimmune vesiculobullous disease with varied presentations. The disease most commonly involves biatoran or bullous mucous membranes; however, involvement of the nasal mucosa, genitilia, anus, esophagus, and larynx has also been reported. MMP has commonly been known as cicatricial pemphigoid, referring to its propensity for scar formation; however, a consensus statement concluded that mucous membrane pemphigoid was the best description for this group of disorders as no previous name adequately encompassed all patients. The pathophysiology of MMP relates to autoantibodies that target subepithelial antigens. The resulting inflammation leads to separation of the epidermal-dermal junction and the formation of subepithelial blisters.

Search Strategy: Literature search included MEDLINE (1966 to July 2008), EMBASE (1980 to July 2008), CENTRAL, Cochrane Database of Systematic Reviews, clinicaltrials.gov, and The National Guideline Clearinghouse databases without language restriction for studies including combinations of the terms mucous membrane, cicatricial, pemphigoid, and epidermolysis bullosa acquisita. Focused searches to help identify laryngeal involvement of MMP included the terms laryn*, pharyn*, throat, glott*, subglott*, supraglott*, hypopharyn*, aytenoids, vocal cord, epiglott*, trach*, airway, and dyspnea. We also scanned the references in the retrieved articles.

Selection Criteria: Inclusion criteria for obtaining the full-text article included original studies, case reports, case series, systematic reviews, meta-analyses, or guidelines reporting on laryngeal involvement in adults of MMP, benign MMP, cicatricial pemphigoid, EBA, or bullous pemphigoid. Epidemiological data, including prevalence, were extracted from articles that included all cases of MMP and did not restrict based on subtype or location. The authors critically appraised the full-text articles using the MOOSE statement for observational studies.

Statistical Analysis: SPSS (version 16.0). Cochrane Mantel-Haenszel tests were used to compare nominal variables. Independent-samples T-test was used to compare means. Kaplan-Meier survival analysis was performed to calculate all-cause survival rates, and the log-rank test was used to compare survival rates among variables.

DISCUSSION & CONCLUSIONS

This systematic review and pooled-data analysis summarizes the available literature on laryngeal MMP. Timely diagnosis of laryngeal involvement is important as it may avoid tracheostomy. Early lesions can be managed medically. Once late-stage scarring occurs, surgical intervention should be considered.