Laryngeal Manifestations of Waldenström's Macroglobulinemia

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INTRODUCTION

Waldenström's macroglobulinemia is a hematologic malignancy that primarily affects the bone marrow and its precursors, with varying extramedullary manifestations. This case report describes our experience with a patient who presented with laryngeal involvement of the hematologic disease. To date, there has been only one prior report in the literature of a patient with such laryngeal involvement, and this patient presented with dysphonia. We present the first review of a patient with laryngeal involvement by Waldenström's macroglobulinemia, highlighting the importance of considering hematologic malignancy in the differential diagnosis of laryngeal mass lesions.

PRESENTATION

An 82-year-old male veteran with a history of Waldenström's macroglobulinemia treated previously with chemotherapy and weekly transfusions as needed presented to the Otolaryngology clinic on an urgent basis for evaluation of airway symptoms. Complaints included several months of dysphonia that had progressed to inspiratory stridor. Evaluation revealed inspiratory stridor, and further questioning in the Otolaryngology clinic revealed several months of mild dysphonia, the onset of which he dated around the time of his initial hematologic diagnosis. Dyspnea had begun one week prior to evaluation, and was initially attributed to his cardiopulmonary comorbidities. Over the four days leading up to evaluation, he noted worsening inspiratory stridor. On examination, the patient was in no acute respiratory distress with normal pulse oximetry values, though he did have inspiratory stridor. Voice was weak and moderately rough. Oral cavity, oropharyngeal, and neck exams were unremarkable. Flexible transnasal laryngopharyngoscopy revealed a large submucosal lesion of the left true vocal cord with subglottal extension causing >90% obstruction of the glottal space, consistent with his Waldenström’s disease. The patient was referred for radiation therapy of this laryngeal lesion, receiving a total dose of 35 Gy in 14 fractions to the larynx. Despite significant improvement in his airway, the patient later died of systemic complications of Waldenström’s prior to decannulation.

MANAGEMENT

Due to the severity nature of his glottic obstruction, the patient was taken urgently to the operating room for awake tracheotomy to secure the airway. Suspension microlaryngoscopy was then performed with the Microlarynx laryngoscope (Medtronic, Minneapolis, MN). Exam revealed a sub mucosal mass of the left true vocal cord, greater than 3cm in maximal diameter and extending to the subglottis. The overlying mucosa was unremarkable. An epithelial cordotomy was made within the subglottic space. The subglottic space was then filled with a saline swab to provide tamponade, and the underlying mass was then excised with preservation of vibratory mucosa and mucosal lining. The patient's postoperative recovery was unremarkable. He tolerated the tracheostomy well and was down-graded to a cuffless tracheostomy tube. Pharyngeal function was substantially improved with no aspiration noted on postoperative videofluoroscopy status with finger occlusion. He was referred for radiation therapy of the larynx, with a field size of 4x4 in 14 fractions of the larynx. Despite significant improvement in his airway, the patient later died of systemic complications of Waldenström's prior to decannulation.

POSTOPERATIVE COURSE

The patient's postoperative recovery was unremarkable. He tolerated the tracheostomy well and was down-graded to a cuffless tracheostomy tube. Pharyngeal function was substantially improved with no aspiration noted on postoperative videofluoroscopy status with finger occlusion. He was referred for radiation therapy of the larynx, with a field size of 4x4 in 14 fractions of the larynx. Despite significant improvement in his airway, the patient later died of systemic complications of Waldenström's prior to decannulation.

HISTOPATHOLOGY

Histopathological evaluation of biopsy specimens demonstrated submucosal atypical polyclonal lymphoid infiltrates consisting predominantly of small, mature-appearing lymphocytes, plasmacytoid lymphocytes and scattered plasma cells with intracytoplasmic Dutcher bodies (Fig. 11). Immunohistochemical stains including Bcl-2, CD20, CD79, CD10, and CD5 were positive for CD20 (Fig. 11), CD79, BCL2, CD23, and Kappa restricted, while CD20 highlighted plasma cells (Fig. 12) in the population. Conal Prolific reds was negative for amyloid. This histological/immunohistochemical examination was consistent with lymphoplasmacytic lymphoma.

REFERENCES


Figure 1: Large submucosal mass at left true vocal cord with subcordal extension causing severe glottic obstruction

Figure 7: Epithelial Cordotomy

Figure 8: Submucosal Flap

Figure 9: Dutcher Bodies

Figure 10: CD20+ immunopositive atypical lymphocytes

Figure 11: CD20+ immunopositive atypical lymphocytes in the submucosal lesion

Figure 12: CD20+ immunopositive atypical lymphocytes in the submucosal lesion

Figure 13: Large submucosal mass at left true vocal cord with subcortical extension causing severe glottic obstruction

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