Primary Natural Killer T Cell Lymphoma of the Supraglottis

Scott H Troob, MD1, David R Friedmann, MD1, Cynthia Liu, MD, PhD2, Maria Suurma MD1
1Department of Otolaryngology – Head & Neck Surgery, New York University School of Medicine, New York, NY 2Department of Surgical Pathology, Hematopathology, and Molecular Pathology, New York University School of Medicine, New York, NY

ABSTRACT

Introduction

Primary hematopoietic neoplasms of the larynx are rare. Secondary tumors are even rarer. The current case of primary laryngeal NK T cell lymphoma is exceptional due to its uniquely protean presentation and aggressive oncologic behavior. The literature review of this case is not comprehensive as the case is an unusual presentation of an unusual location for an aggressive subtype of extranodal lymphoma. The case report and literature review should aid in the differential diagnosis.

Methods

The patient’s medical records were reviewed, including the clinical history, laboratory, radiologic, and pathologic data. Additionally, a literature search was performed for cases of laryngeal lymphoma, with emphasis on NK T cell lymphomas.

Results - Case Presentation

A 22-year old male tombstone carver from El Salvador, who immigrated to the United States two years earlier, presented to the Bellevue Emergency room complaining of two months of cough, hemoptysis, fever, dysphagia, odynophagia, hoarseness, night sweats and a 70-pound weight loss. Work up for tuberculosis, HIV and serologies for systemic granulomatous conditions were negative. Otolaryngologic evaluation demonstrated normal-appearing nasal mucosa and oral cavity. Laryngoscopy revealed a thickened epiglottis with polypoid mucosa of the vallecula and the supraglottis. Biopsy of the epiglottis demonstrated NK T cell lymphoma, nasal-type with CD3 and CD56 positivity and histologic features of an aggressive EBV-associated form. Imaging and bone marrow biopsy confirmed its confinement to the supraglottis. He is currently undergoing concurrent chemoradiation therapy.

Discussion

NK T cell lymphoma, formerly known as Kibb linalo granulomas or angiocentric lymphoma, is an extranodal lymphoma that may present in the head and neck. It most often occurs in the parenchymal sinususes and nasal cavity of young to middle age men. As seen in our case, tissue necrosis with pseudoepitheliomatous hyperplasia is thought to be secondary to perivascular invasion and vascular occlusion. Unlike the classic presentation, our patient did not have involvement of the nasal cavity or paranasal sinus.

Conclusions

Extensive involvement occurs in approximately 25% of lymphomas. When such involvement occurs in the larynx, it most often develops in the lymphoid collections in the lamina propria and ventricles of the supraglottic region. A recent review of primary laryngeal lymphoma of the larynx places the mean age of onset in the 7th decade of life (range 4-81 years). Patients often present with complaints of hoarseness, cough, dysphagia, globus sensation, and/or weight loss and fever are also common.

Radiographically, laryngeal lymphomas demonstrate homogenously enhancing lesions that may result in thickening of the epiglottis with submucosal enhancement, air in retropharyngeal space, thickening of epiglottis with submucosal enhancement, and uninvolved glottis and subglottis.

References


Primary laryngeal NK T cell lymphoma is a rare, aggressive subtype of extranodal lymphoma with a unique protean presentation and aggressive oncologic behavior. The literature review of this case is not comprehensive as the case is an unusual presentation of an unusual location for an aggressive subtype of extranodal lymphoma. The case report and literature review should aid in the differential diagnosis.

Results - Case Presentation

A 22-year old male tombstone carver from El Salvador, who immigrated to the United States two years earlier, presented to the Bellevue Emergency room complaining of two months of cough, hemoptysis, fever, dysphagia, odynophagia, hoarseness, night sweats and a 70-pound weight loss.

On initial fiberoptic examination in the ER the patient's nasal cavity was appreciated. When such involvement occurs in the larynx, it most often develops in the lymphoid collections in the lamina propria and ventricles of the supraglottic region. A recent review of primary laryngeal lymphoma places the mean age of onset in the 7th decade of life (range 4-81 years). Patients often present with complaints of hoarseness, cough, dysphagia, globus sensation, and/or weight loss and fever are also common.

The patient received 6 weeks of radiation therapy and 3 cycles of chemotherapy - DeVIC (dexamethasone, VP16, ifosfamide, carboplatin). At 6 months follow up, he is clinically and radiographically without evidence of disease.

CONCLUSIONS

Lymphoma of the larynx represents 1% of malignant laryngeal tumors. Primary laryngeal NK T cell lymphoma with disease isolated to the larynx is exceptionally rare. Medical work up assists in ruling out infectious or inflammatory lesions. Radiographic data, directed biopsies, and serologies for systemic granulomatous conditions (serum ACE, C-ANCA, P-ANCA) were all negative. Medical work up to rule out infectious or inflammatory lesions along with directed biopsies should help in the diagnosis.

REFERENCES