Case Report

Mucosa-Associated Lymphoid Tissue Lymphoma of the Larynx: Rare but Existing Entity

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Abstract

Mucosa-associated lymphoid tissue (MALT) lymphomas are a subtype of non-Hodgkin lymphoma stemming from marginal zone B-cells. In this case report, we present one patient with an extremely rare localization of MALT lymphoma to the larynx.

INTRODUCTION

Extra nodal lymphoma involving the larynx is exceedingly rare, accounting for less than 1% of all primary laryngeal neoplasms [1]. Most lymphomas involving the larynx involve other sites as well, including the salivary glands, thyroid, nasopharynx, and tonsils. Recognition of the clinical presentation of laryngeal lymphoma may prevent inappropriate management, particularly if definitive surgical decisions are made on the basis of frozen sections. The treatment of laryngeal lymphoma differs from that of other submucosal lesions such as laryngeal cysts, neurofibromas, lipomas, myxolipomas, hemangiopericytomas, paragangliomas, laryngeal amyloidosis, neurilemomas, and Teflon granulomas [2]. Whereas these other lesions generally require resection, resection is contraindicated in laryngeal lymphoma.

We report this case of laryngeal lymphoma because, despite its relative rarity, the consequences of a missed diagnosis warrant awareness of and vigilance for this type of laryngeal tumor.

CASE PRESENTATION

Extranodal laryngeal lymphoma is extremely rare. We report a case of primary laryngeal lymphoma in a 48-year-old woman presented with 6 months of worsening throat discomfort, intermittent cough, and dyspnea with exertion. She denied any hemoptysis, otalgia, or odynophagia. She had no other significant past medical or social history including no history of tobacco use.

A CT neck with contrast demonstrated a mass involving the anterolateral right subglottic larynx with extension along the upper cervical trachea (Figure 1).

In-office flexible nasolaryngoscopy identified a fleshy pink-colored mass in the immediate right subglottis obstructing approximately 50% of the airway. She then underwent microdirect laryngoscopy and bronchoscopy for debulking and biopsy (Figure 2).

Operative findings were notable for extension of the mass approximately 1 cm below the true vocal fold.

Pathologic analysis noted small B-cells positive for CD19, CD20, and PAX5 and negative for CD5, CD10, and cyclin D21. FISH was negative for the 18q21 MALT1 gene. The combination of these findings confirmed the diagnosis of extranodal marginal zone B-cell lymphoma of MALT type was made (Figure 3).

Figure 1 Axial CT neck with contrast demonstrating a right subglottic mass.
DISCUSSION

Lymphoma is one of the most common malignancies of the head and neck, second only to squamous cell carcinoma. Extranodal tumors in the head and neck are usually non-Hodgkin’s lymphomas [3,4]. The larynx appears to be a rare site of extranodal lymphoma, accounting for less than 1% of all primary laryngeal neoplasms [1]. By 1976, only 14 cases had been reported in the English-language literature [1]. By 1989, 11 more cases of primary laryngeal non-Hodgkin’s lymphoma had been added (5, 20). As of this writing, the total number of reported cases was approaching 90 [5]. Our review of the literature also revealed that the median age of patients with laryngeal lymphoma was 60 years (range: 14 to 81). The distribution between males and females was almost equal. The most common symptom at presentation was hoarseness, which had been present from 2 to 18 months. Other reported symptoms were dysphonia, dysphagia, stridor, and cough. Although these symptoms are indistinguishable from those of other laryngeal tumors, the macroscopic appearance of non-Hodgkin’s lymphoma of the larynx may raise the suspicion of an attentive clinician. Most laryngeal lymphomas present as a submucosal mass or a polypoid tumor; they are smooth, non-ulcerated, and Gray-white. They are usually located in the supraglottic region, and they have a particular tendency to involve the aryepiglottic folds, although some cases have been reported in the subglottis. While suggestive, none of these features is pathognomonic for a laryngeal lymphoma. Definitive diagnosis depends on histologic examination of a biopsy specimen. Primary laryngeal lymphomas probably arise from specialized submucosal aggregates of lymphoid cells present in the lamina propria of the supraglottic area and epiglottis [6-8]. Tumor growth slowly expands the overlying mucosa, which remains intact, and these results in a benign-appearing mass. Squamous cell carcinomas, in contrast, arise in the squamous epithelium and present as an irregularity involving the free margin of the laryngeal structures. A wide spectrum of histologic subtypes of laryngeal lymphomas has been reported. The great majority of laryngeal non-Hodgkin’s lymphomas have been of B-cell lineage; very few T-cell immunophenotypes have been reported [9,10]. Using the working classification of these tumors, a high proportion were diffuse large-cell lymphomas [11,12]. Historically, radiotherapy has been the primary modality of therapy for these tumors. Results have been fairly good, and long follow-ups have found few recurrences [13-15]. Considering the systemic nature of most cases of non-Hodgkin’s lymphoma, we believe that chemotherapy has a role, especially in cases of low-grade lymphoma. A definitive diagnosis continues to rely on histologic examination of a biopsy specimen. Care in decision making should be exercised regarding frozen sections. There is little role for surgical resection.

CONCLUSION

This case report demonstrates that MALT lymphoma can present with much more benign and subtle symptoms. This highlights the importance of clinicians to keep broad differentials and consider MALT lymphomas in the setting of laryngeal masses.

CONSENT

Informed consent was obtained from all participants included in the study.

REFERENCES

1. Aster J, Freedman A. Clinical manifestations, pathologic features, and diagnosis of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT)-up to date. 2019.


