Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma Presenting as Subglottic Stenosis: Single-Agent Treatment Using Rituximab
Clifford Bielinski, Hung S. Luu and Ted Mau

Otolaryngology -- Head and Neck Surgery 2014 150: 334 originally published online 13 December 2013
DOI: 10.1177/0194599813515632

The online version of this article can be found at:
http://oto.sagepub.com/content/150/2/334

Published by:
SAGE
http://www.sagepublications.com

On behalf of:
AMERICAN ACADEMY OF
OTOLARYNGOLOGY--
HEAD AND NECK SURGERY

American Academy of Otolaryngology- Head and Neck Surgery

Additional services and information for Otolaryngology -- Head and Neck Surgery can be found at:

Email Alerts: http://oto.sagepub.com/cgi/alerts
Subscriptions: http://oto.sagepub.com/subscriptions
Reprints: http://www.sagepub.com/journalsReprints.nav
Permissions: http://www.sagepub.com/journalsPermissions.nav

>> Version of Record - Feb 3, 2014
OnlineFirst Version of Record - Dec 13, 2013
What is This?
Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma Presenting as Subglottic Stenosis: Single-Agent Treatment Using Rituximab

Clifford Bielinski¹, Hung S. Luu, MD², and Ted Mau, MD, PhD¹

Keywords
MALT lymphoma, laryngeal lymphoma, subglottic stenosis, rituximab

Introduction
Inspiratory stridor due to subglottic stenosis is a well-known entity, with the vast majority of adult cases due to intubation, external trauma, and less commonly idiopathic progressive subglottic stenosis (IPSS) and systemic rheumatic diseases. With very few exceptions, the widening of airway caliber requires surgical intervention. We present a case with this relatively common presentation but caused by an exceedingly rare disease entity, a laryngeal MALT lymphoma treated successfully with single-agent chemotherapy.

Case Report
An otherwise healthy 88-year-old female with no history of intubation developed progressive inspiratory stridor over a 2-year period. She was initially diagnosed with chronic obstructive pulmonary disease (COPD) and was treated empirically without relief. A computed tomography (CT) scan of her neck and chest with contrast identified subglottic stenosis as the only radiographic abnormality. Flexible laryngoscopy showed circumferential narrowing of the subglottic airway (Figure 1A). Since the stridor had become more severe, she underwent microlaryngoscopy with the intent of biopsy and radial incisions with dilation, under general anesthesia with jet ventilation. Intraoperatively, it was noted that the area of stenosis was covered by relatively normal-appearing mucosa (Figure 1B), in contrast with IPSS, in which the mucosa is invariably inflamed. Palpation revealed the subglottic walls to have a spongy, rather than firm, consistency. A large biopsy on the left subglottal wall was taken. More aggressive resection was not performed in order to avoid cicatrix formation. A tracheotomy was not required. The specimen showed a dense lymphoid infiltrate of predominantly small lymphocytes. These positively labeled for CD20 and negatively for CD5, CD10, and CD43 on immunohistochemistry (Figure 2). Immunoglobulin gene rearrangement study by polymerase chain reaction (PCR) was positive for a monoclonal B-cell population. Taken together, these results were consistent with a low-grade B-cell, extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). Six weeks following the biopsy and after the diagnosis was confirmed, the patient was started on rituximab (Rituxan) therapy, with 4 weekly infusions without complication. Her breathing was improved following treatment, and exam showed a widened subglottis. She remained...
asymptomatic at 18 months follow-up and continues to be in remission.

Exemption from review was granted by the University of Texas Southwestern Institutional Review Board.

Discussion

This case is notable for 2 features. First, it highlights an exceedingly rare disease entity with a relatively common presentation, namely, stridor associated with subglottic stenosis. The top differential diagnoses based on her history were IPSS and rheumatic disease such as Wegener’s granulomatosis. Her advanced age, however, did not fit the profile of disease onset for either condition. A new disease at such an advanced age would favor a neoplastic process, yet her exam had a benign appearance and belied the unusual malignancy.

MALT lymphoma is an indolent B-cell malignancy and a subtype of non-Hodgkin’s lymphoma whose optimal management remains elusive. It occurs most commonly in the gastrointestinal tract but has also been described in a variety of extranodal sites including the respiratory tract, lacrimal and salivary glands, thyroid, and breast. Primary lymphomas of the larynx account for less than 1% of laryngeal malignancies. Isolated MALT lymphomas of the larynx are exceedingly rare. Our literature search uncovered 26 cases described in 24 case reports, with the supraglottis the predominant subsite. Only 4 cases were localized to the subglottis.

The second notable feature of this case is its management. As a consequence of their rarity, there are no guidelines on the best treatment for MALT lymphomas of the larynx. The majority of cases reported were treated with surgical resection, radiotherapy, conventional chemotheraphy, or a combination thereof. Three cases were treated with a chemotherapy regimen that included alkylating agents plus rituximab, an anti-CD20 monoclonal antibody that targets B-cell non-Hodgkin’s lymphomas. To our knowledge, the current report is the first to illustrate the successful single-agent treatment of a laryngeal MALT lymphoma using rituximab. Unlike conventional chemotherapy drugs, rituximab has a cytotoxic effect that is highly specific to B-cells and is therefore better tolerated with fewer side effects. Our result suggests the possibility of treating isolated MALT lymphoma of the larynx without the use of the more toxic agents and without surgery or radiotherapy. This treatment approach avoids postsurgical scarring and mucosal scarring from radiotherapy, both important considerations for interventions on the subglottic airway.

Author Contributions

Clifford Bielinski, data acquisition, manuscript writing, final approval; Hung S. Luu, data acquisition, manuscript revision, final approval; Ted Mau, conception, data acquisition, manuscript writing, final approval.

Disclosures

Competing interests: None.
Sponsorships: None.
Funding source: None.

References