Inflammatory Myofibroflastic Tumor of the Larynx: Case Report and Review of the Literature

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Abstract

Objective: Describe a case of Inflammatory Myofibroblastic Tumor (IMFT) of the larynx, analyze the literature regarding the disease process, and discuss prognosis.

Methods: Retrospective review of a single case of IMFT involving the subglottic region at the University of Texas Medical Branch.

Results: We present IMFT of the larynx, a benign condition of which only 28 cases in the larynx have been described. Our patient's tumor was ALK-1 positive, indicating a favorable prognosis. Microsuspension laryngoscopy with laser excision has been the mainstay of treatment, with or without steroid injection.

Conclusion: IMFT is a benign tumor of the larynx, and conservative excision is adequate for tumor control. Thorough physical examination, a wide differential diagnosis, and pathological analysis are important to prevent overtreatment and to make the appropriate diagnosis.

Introduction

Inflammatory myofibroblastic tumors (IMFTs) are rare neoplasms of unclear origin that are most commonly found in the lungs and the abdominopelvic region, with rare involvement of the larynx.[1, 4, 5] Some cases of IMFT have shown immunohistochemical expression of anaplastic lymphoma kinase (ALK), which confers a higher likelihood recurrence but a lower chance of metastasis and therefore is prognostically pertinent.[5] In this report we wish to add to the body of literature on this disease, given the uncommon findings of ALK positivity and subglottic extension.

Case Report

A 23-year-old female presented to the laryngology clinic with a 5-month history of fluctuating hoarseness. She described a normal voice each morning, but experienced vocal fatigue with use as the day progressed. Her review of systems was otherwise unremarkable.

Videostroboscopic examination revealed a cystic lesion involving the inferior edge of the middle third of the right true vocal cord with extension into the subglottic region (**Figure 1**). Examination also revealed signs of laryngeal reflux. The patient refused surgical removal and opted for medical treatment of reflux.

At two-months follow up, despite partial compliance with reflux therapy, she had no significant improvement in her voice. She was scheduled for microsuspension laryngoscopy with mass excision. The firm, sessile, papillomatous mass was completely removed from the glottis using CO_2 laser.

Gross analysis of the surgical specimen showed a tan-pink, soft tissue mass measuring 0.7 x 0.4 x 0.3 cm. Histopathological analysis demonstrated that the specimen was an inflammatory myofibroblastic tumor, with a spindle cell component that was immunohistochemically positive for SMA and ALK-1 (**Figure 2**).

Discussion

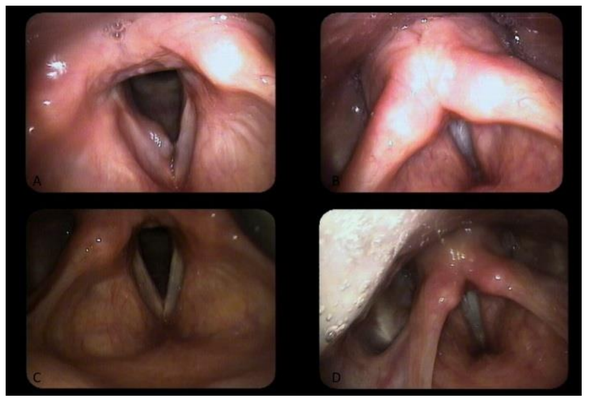
Laryngeal IMFTs are rare, benign, inflammatory masses, of which 28 have been previously reported in the English literature. Of these 28, only 7 originated in the subglottic area.[2]

The most commonly reported symptom of IMFT of the larynx is hoarseness, but dysphonia, stridor, shortness of breath, and globus sensation are also common.[1, 4] In the case presented, the patient reported hoarseness for five months. We emphasize the importance of the use of videostroboscopic examination as well as histological examination of surgical specimen to rule out malignancy.

The *ALK* gene normally encodes a receptor tyrosine kinase that is a part of the insulin growth factor receptor family. IMFTs positive for ALK have been deemed to have a favorable prognosis, with rare malignant transformation. Metastasis of the lesion has only been found in ALK-negative subsets. Alternatively, ALK-positive lesions have been shown to have higher rates of recurrence, supporting the theory that these lesions are neoplastic, yet still benign.[5] The IMFT removed from our patient was ALK-positive, indicating a need for follow up to monitor for recurrence.

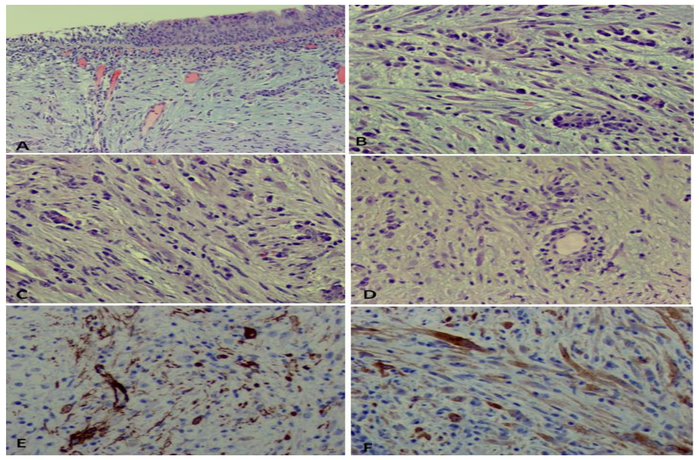
While laryngeal IMFTs are rare neoplasms, it is important to recognize them based on clinical history, picture, and appropriate physical exam, as their symptoms can mimic other diseases processes, and their structure can mimic that of malignant neoplasm. Methods for the removal of laryngeal IMFTs include laser or surgical excision, with or without postoperative steroid therapy.[2] Astute pathologists suspecting an IMFT of the larynx should stain for ALK, as ALK positivity indicates a low chance of metastasis, but a higher likelihood of recurrence, both of which are important prognostic pieces of information.

Figure 1



(A, B) Videostroboscopic examination showing a cystic lesion involving the inferior edge of the middle third of the right true vocal cord with extension into the subglottic region.
(C, D) Videostroboscopic examination at first postoperative visit, showing no recurrence.

Figure 2



The submucosal lesion is composed of spindle cells with myxoid background, and the surface is lined with pseudostratified respiratory epithelium suggestive of anatomic location of false vocal cord (A). The spindle and elongated nuclei with eosinophilic cytoplasm (B and C) are noted with prominent lymphoplasmacytic infiltrate, especially around the small vascular channels (B and D). Mitosis is rare. The spindle cell components are focally positive for smooth muscle actin (E) and diffusely positive for Alk-1 (F).

References

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