Inflammatory Myofibroblastic Tumor of the Larynx—A Case Report

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Summary: Objectives. Inflammatory myofibroblastic tumor (IMT) is a borderline neoplasm with uncertain malignant potential. It is a rare disease also referred to as an inflammatory pseudotumor, a plasma cell granuloma, and an inflammatory fibrosarcoma. IMT rarely also involves the head and neck region with only 50 cases of laryngeal IMT reported in the literature, and this is the first case with reported magnetic resonance imaging (MRI) findings.
Methods. A 37-year-old man with a 1-year history of hoarseness, dysphagia, and fatigue presented with a right vocal fold submucosal mass and was treated conservatively.
Results. The MRI of the neck revealed a mildly spontaneously hyperintense right true vocal fold on GRE images and relative hyperintensity on fat-saturation T2-weighted images. A biopsy of the right-sided submucosal laryngeal mass was performed and the pathologic examination revealed a lesion consistent with an IMT.
Conclusion. IMT is a borderline neoplasm with uncertain malignant potential. There are many variants of IMT and its etiology is not truly understood. In general, IMT of the larynx has a benign clinical course with low rates of recurrence.
Key Words: Laryngology–Inflammatory myofibroblastic tumor–Vocal fold tumor–Larynx.

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare borderline neoplasm with uncertain behavior. It is also known as inflammatory pseudotumor, plasma cell granuloma, and sometimes referred as inflammatory fibrosarcoma with each having some variants reported in the literature.[1] IMT mainly affects the lungs, the mesentery, and the omentum. Rarely does it affect the head and neck region. To date and to the author's knowledge, less than 50 cases of IMT of the larynx have been reported in the English literature.

In this article, a case of a 37-year-old man with IMT of the larynx is presented and a discussion on the clinical presentation of IMT, the histopathology, as well as the management will follow.

CASE REPORT

The presentation

A 37-year-old man was seen in our Otolaryngology clinic with a 1-year history of hoarseness, dysphagia, and fatigue. He was a nonsmoker and did not drink alcohol. He was known for a vitamin B12 deficiency and gastro-oesophageal reflux. His hoarseness was rough, effortful with fatigue. He did not have difficulty eating or drinking but described an increasingly cumbersome swallow during meals. At that time, he was also being investigated for left-sided body weakness. His general head and neck examination was normal. Flexible transnasal pharyngolaryngoscopy revealed a right vocal fold submucosal mass, with generalized thickening along the entire length of the fold (Figure 1). The fold did exhibit normal mobility. The mucosal wave was absent on the right fold and diminished anteriorly on the left fold due to the bulkiness of the lesion. Mild signs of laryngopharyngeal reflux were also present.

Radiology. The magnetic resonance imaging (MRI) of the neck revealed a mildly spontaneously hyperintense right true vocal fold on fat saturated T1 gradient-echo (GRE) images and relative hyperintensity on fat-saturation T2-weighted images (Figures 2–5). It demonstrated strong enhancement following contrast infusion. The thickening of the right true vocal fold extended to the region of the anterior commissure but did not cross it. There was bulging of the fold into the subglottic space, but there was no true extension. The barium swallow and the modified barium swallow were unremarkable.

Surgery. Suspension microlaryngoscopy and cold steel biopsy of the right-sided submucosal laryngeal mass was performed. A lateral microflap was performed and a submucosal plane was lifted. The mass was extremely rigid, most comparable with wood consistency. During the debridement, the surgeon took great care not to harm the mucosa overlying the lesion.

Pathology

Biopsies of the mass showed a low-grade myofibroblastic tumor without marked atypia, necrosis, or mitoses admixed with lymphocytes and plasma cells (Figure 6). There was no evidence of definite malignancy. The immunochemistry stains were positive for smooth muscle actin (SMA), desmin, and CD34. They were negative for CKAE1/AE3, myogenin, caldesmon, beta-catenin, S100, and anaplastic lymphoma kinase (ALK) protein. ALK protein fluorescence *in situ* hybridization was also negative. Moreover, the Ki67 proliferative index was low.

Management

Postoperatively, the patient was reviewed in our institutional tumor board conference. The team agreed for a conservative

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Consent was obtained by the patient after consultation with our hospitals Research Ethics Committee and thus formal institutional review board was not required.
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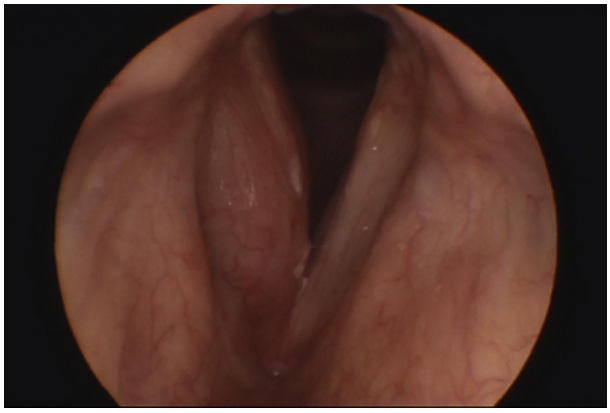

FIGURE 1. Flexible transnasal pharyngolaryngoscopy revealing a right vocal fold submucosal mass with generalized thickening of the entire length of the fold.

approach, which included laser debulking in the event of voice or airway compromise with close observation with serial MRI.

There was a slight increase of the mass at a follow-up appointment clinically a few months postoperatively, without fear of airway obstruction. A trial of oral steroids for 4 weeks was initiated. Fifty milligrams of prednisone were given daily for 1 week, and the doses were tapered over the next 3 weeks. This resulted in a significant reduction in the tumor size. Currently, the patient is stable for 6 months, has a voice quality that is manageable, and a patent airway.

DISCUSSION

IMT is classified as a borderline neoplasm with uncertain behavior. Although, IMT of the head and neck is rare, the larynx is the most common site in the latter.[2,3]

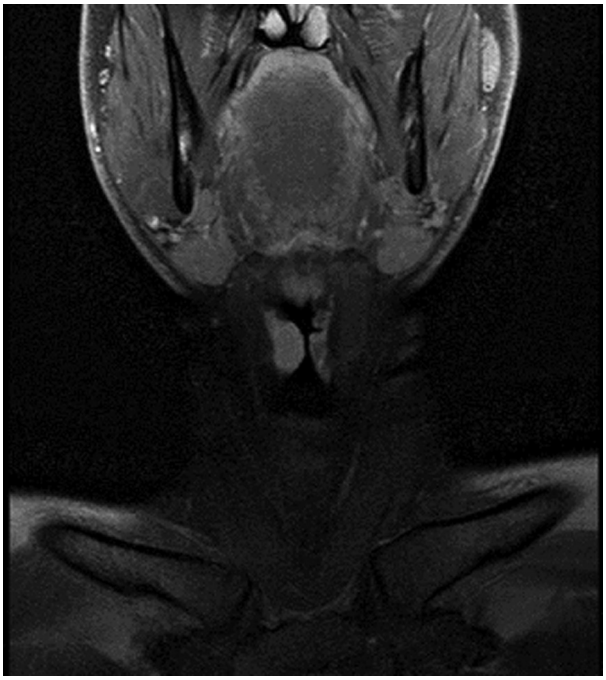

FIGURE 2. MRI Coronal T1W postcontrast showing the thickening of the right true vocal fold.

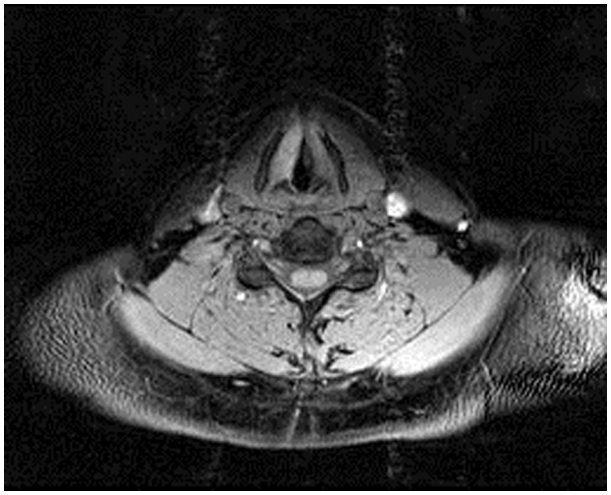

FIGURE 3. MRI GRE precontrast of the true vocal folds.

The etiology of IMT is still unclear. It was first thought to be of inflammatory origin; however, researchers refuted this hypothesis by finding clonal cytogenetic aberrations that would explain the behavior of this tumor.[4] From the literature, common factors predisposing to IMT, such as trauma, smoking, and immune responses, have been proposed.[5]

The mean age of patients with IMT is around 43 years. They most often present to the clinic with hoarseness or dysphonia. Other symptoms that are occasionally present in this disease are dyspnea, stridor, globus, dysphagia, hemoptysis, cough, and otalgia.[5] A small portion of patients with IMT can also have a syndrome of fever, weight loss, growth failure, malaise, anemia, thrombocytosis, polyclonal hyperglobulinemia, and elevated erythrocyte sedimentation rate, but these have not been documented in laryngeal IMT cases.[4] IMT of the larynx is most often localized in the vocal folds, in the subglottis, or in the aryepiglottic folds.[5]

Our presentation of the IMT was particular on imaging. The use of MRI has not been reported in cases of laryngeal IMT. The

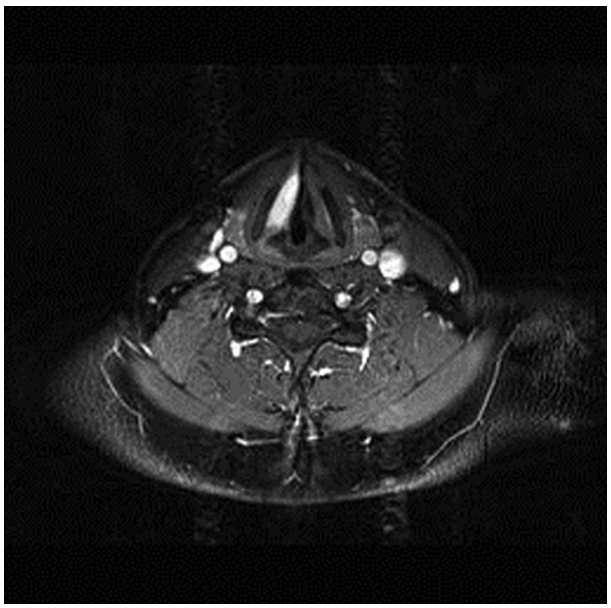

FIGURE 4. MRI GRE postcontrast of the true vocal folds.