Inflammatory Myofibroblastic Tumor of the Epiglottis Excised with CO₂ Laser

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INTRODUCTION

The inflammatory myofibroblastic tumor (IMT) is a rare and distinctive neoplasm with intermediate biologic potential. IMTs have a predilection for the abdominopelvic region and lungs of children and young adults, though virtually any site in the body can be affected. IMT’s involving the larynx are rare, with less than 40 cases reported in the English literature. To our knowledge, this is the first reported case of an IMT localized to the epiglottis.

CASE REPORT

A 66-year-old female presented with complaints of dysphonia and odynophagia having progressively worsened over several weeks. She denied any other complaints and her past medical history was non-contributory. A flexible fiber-optic laryngoscopy (FFL) revealed a small granular lesion with surrounding erythema involving the tip of the epiglottis (Figure 1). The remainder of her exam was unremarkable. A computed tomography (CT) scan of the neck with contrast displayed a thickened suprahyoid epiglottis bilaterally. The patient subsequently underwent micro-direct suspension laryngoscopy (MDSL) with wide and complete excision of the epiglottic lesion with CO₂ laser. No adjuvant therapy was used. Macropscopic analysis revealed an excised 3.2 x 1.7 x 0.5 cm epithelial specimen with a 1.2 x 0.7 cm depressed granular lesion with fragments of pink, red, and tan tissue on the mucosal surface. Biopsies of the mass showed ulcerated mucosa with a subepithelial collection of epithelioid and spindled histiocytes admixed with lymphocytes and plasma cells (Figure 2). The patient experienced resolution of her dysphonia as early as 4 weeks postoperatively and showed no evidence of recurrence after two years of follow-up.

DISCUSSION

Inflammatory myofibroblastic tumor (IMT) is defined by the World Health Organization as an intermediate potential soft-tissue tumor characterized by myofibroblastic spindle cells on a background of inflammatory cells. Multiple pseudonyms having been used to describe the same group of lesions, including inflammatory pseudotumor, histiocytoma, and plasma cell granuloma. IMTs frequently occur in the lung but rarely involve the head and neck region. Laryngeal occurrence is rare, with approximately 40 cases having been described in the English literature. The most common sites of laryngeal IMT are the glottis (51%) and subglottis (22.5%). This is the first reported case of an IMT of the epiglottis.

Laryngeal IMTs tend to affect adult (mean age, 44.5 years) males (71%). The etiology of IMTs remains unclear, though some suspect an immunologic response to stimuli including microorganisms, tissue damage, smoking, foreign bodies, or neoplasms.

IMTs may present with non-specific findings including pain, fever, and weight loss. Patients with laryngeal IMT commonly present with dysphonia (74%), as did our case. Less often they present with stridor (29%), dyspnea (22.5%), and globus (16%).

Histologically, IMTs are composed of myofibroblasts mixed with an infiltrate of inflammatory cells (lymphocytes, plasma cells, eosinophils, and macrophages). Three basic histologic patterns described by Coffin et al. are universally accepted:

(a) myxoid, vascular, and inflammatory areas resembling nodular fasciitis;
(b) compact spindle cells with intermingled inflammatory cells resembling fibrous histiocytoma; and
(c) dense plate-like collagen resembling a desmoid or scar. Highly atypical cells are seen in rare IMTs which undergo histologic malignant transformation. Our patient’s biopsies revealed ulcerated mucosa with a subepithelial collection of epithelioid and spindled histiocytes admixed with lymphocytes and plasma cells. There was no atypia or necrosis. These findings are consistent with the above-mentioned type (b) IMT. Immunohistochemical staining can aid in the diagnosis of laryngeal IMT. Muscle specific actin, vimentin, and smooth muscle actin often present in IMT, though specificity is low.

Though the radiologic findings of epiglottic thickening in our case were rather non-specific, a contrast-enhanced CT scan displaying a well-enhanced mass can assist the diagnosis. Magnetic resonance imaging will show a well-enhanced lesion on gadolinium-enhanced T1-weighted images.

The differential diagnosis includes soft tissue tumor such as angioleiomyoma, leiomyoblastoma, or leiomyosarcoma and malignant tumor such as squamous cell carcinoma, lymphoma, malignant fibrous histiocytoma, or spindle cell squamous carcinoma.

The mainstay of treatment for laryngeal IMT is complete surgical excision (with or without laser). MSDL or direct laryngoscopy with endoscopy is preferred over open technique, which is reserved for recurrence, poor visualization, or when malignancy cannot be excluded. Steroids and non-steroidal anti-inflammatory drugs have been used independent and as adjuvant therapy, though individual therapy seems to have less efficacy. The role of antibiotics is less certain. Radiation and chemotherapy are used to treat recurrent IMTs and lesions that have undergone malignant transformation. In some patients with multiple recurrences, a laryngectomy may be warranted. REGARDLESS the treatment used, a high recurrence rate (27.3%) for laryngeal IMTs warrants thorough postoperative follow-up. No cases of laryngeal IMT have reported metastases to date.

Our patient underwent successful excision of an epiglottic IMT with CO₂ laser and showed no evidence of recurrence after two years of follow-up.

CONCLUSION

Laryngeal IMTs represent a rare disease and present a diagnostic challenge. IMTs clinically appear suspicious and histologically may mimic malignancy. The benign nature of this lesion precludes aggressive treatment; however, IMT’s potential for local expansion warrants prompt intervention. Complete endoscopic surgical excision (with or without adjuvant therapy) is the preferred treatment for laryngeal IMTs. This is the first known report of a laryngeal IMT affecting the epiglottis. Otolaryngologists should be aware of these rare entities, their clinical presentation, and how they are successfully managed.

REFERENCES