INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE LARYNX

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Abstract: Background. Inflammatory myofibroblastic tumor, composed of myofibroblastic spindle cells with acute and chronic inflammatory cells, is an unusual, benign solid mass that mimics a neoplastic process.

Methods. We report a rare case of a patient with a laryngeal inflammatory myofibroblastic tumor. Laryngoscopy demonstrated a submucosal mass involving the right false cord. The mass was a well-enhanced supraglottic lesion on CT scan. It showed medially high signal intensity and peripherally low signal intensity on T2-weighted MR images, and it displayed a high magnetization transfer ratio; before surgery, it was believed to be a malignant tumor. Laryngoscopic biopsy was performed. Pathologic features of the specimen were diagnostic for inflammatory myofibroblastic tumor.

Results. Steroid therapy was chosen for further treatment. No recurrence was observed for 4 years.

Conclusion. In patients with chronic hoarseness who have a malignant-looking submucosal laryngeal mass, inflammatory myofibroblastic tumor should be considered. Conservative surgery and steroid treatment are advocated because of laryngeal preservation.

Keywords: larynx; inflammatory myofibroblastic tumor; CT imaging; MR imaging; steroid therapy

Inflammatory myofibroblastic tumor (IMFT) is a benign fibroinflammatory mass of an unknown origin, and the true nature of IMFT is only now beginning to be revealed.1 IMFT has many synonyms: plasma cell granuloma, inflammatory pseudotumor, xanthogranuloma, histiocytoma, and myofibrohistiocytic proliferation.2 The lung, gastrointestinal tract, and orbit are the most common sites for IMFT. Laryngeal IMFT is extremely rare,3 and it may easily be misinterpreted as a malignant tumor because of its clinical findings, severe symptoms, and rapid proliferation. We present here a case of a malignant-looking lesion found in the larynx that was pathologically confirmed as an IMFT in a 52-year-old female patient. We also review the clinical and pathologic aspects of this unusual disease entity.

CASE REPORT

A 52-year-old woman was seen with a 1-year history of voice change. No other constitutional symptoms were present nor was there a history of any trauma or previous operation. She was a non-smoker. The laryngoscopic examination demon-
strated a contour bulging firm glistening submucosal mass involving the right false cord. Contrast-enhanced axial CT scan showed a well-enhancing, elongated mass involved the right supraglottic area (Figure 1A), and there was no lymphadenopathy larger than 1 cm at the maximal diameter and no lymph nodes with internal necrotic foci. On MRI, the lesion was isointense, with the uninvolved side of the larynx on T1-weighted images and the contrast-enhanced study demonstrating a well-enhanced mass. On T2-weighted images, the lateral portion of the mass showed relatively low signal intensity, and the medial portion revealed high signal intensity (Figure 1B). Magnetization transfer (MT) images were also obtained. Regarding the MT pulse, it was a Gaussian pulse (pulse duration of 7 msec, maximum transmitter power, and equivalent flip angle of 500 degrees). It was off resonant by 2 kHz. On application of MT, the lesion showed suppressed signal intensity and the magnetization transfer ratio (MTR: 1-[signal intensity of the MT image/signal intensity of the pre-MT image]) of the tumor was 0.347. The higher the MTR value for the lesion, the greater the degree of suppression of the signal intensity after the MT pulse and the more macromolecular cell wall protein there is in the lesion; such a signal is considered to be from a malignant tumor with high cellularity. We performed a laryngoscopic biopsy for the evaluation of the lesion. Histologic examination of the surgical specimens demonstrated multifocal infiltrations of lymphocytes, histiocytes, plasma cells, and eosinophils with occasional lymphoid follicles. Between the inflammatory cells, spindle cells with eosinophilic cytoplasm and oval to elongated nuclei were seen (Figure 2). These cells were positive for vimentin and smooth muscle actin but negative for S-100 protein and desmin. The lymphoid cells were mixture of CD20-positive B cells and CD3-positive T cell. The special stains

![Figure 1. Axial contrast-enhanced CT scan (A) during admission shows a relatively well-enhancing, elongated mass centered in the right supraglottic larynx (arrows and arrowhead). Corresponding axial T2-weighted MR image (B) shows a heterogeneous mass involving the right supraglottic larynx. The lesion mainly exhibits a high signal intensity (arrowhead). Note the prominent low signal intensity of the anterolateral portion of the mass (arrows).](image1)

![Figure 2. Histologically, the tumor is composed of spindle-shaped myofibroblasts in collagenous and inflammatory background (hematoxylin-eosin stain, original magnification ×200). [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.](image2)
were negative for acid-fast bacillus and fungi. The morphologic and immunohistochemical studies were diagnostic for IMFT. High-dose corticosteroid therapy was chosen for further treatment. The patient’s clinical status gradually improved, and follow-up at 4 years reveals no evidence of recurrence.

**DISCUSSION**

IMFT, which has been also referred to as inflammatory pseudotumor (IPT), plasma cell granuloma, and pseudosarcomatous myofibroblastic lesion, manifests as a space-occupying lesion composed of a benign lymphoplasmacytic and myofibroblastic infiltrate. In the head and neck, IMFT most often affects the periorbital and orbital soft tissue. There have been limited cases reported involving the sinonasal cavity, nasopharynx, larynx, oral cavity, salivary gland, temporal bone, lymph nodes, pterygomaxillary space, parapharyngeal space, and skull base.1–5 Although its histopathologic nature is benign, it may be difficult to differentiate this lesion from a malignant tumor because of its local invasiveness and its tendency to recur. It is important to recognize the imaging features of IMFT, because it can potentially mimic multiple clinical entities.

The etiology and pathogenesis of IMFT still remains a mystery. Some investigators consider it an immunologic host response to many different stimuli, including infectious agents, microorganisms, adjacent necrotic tissue, neoplasms, foreign bodies, and some kinds of tissue injury.6 Cytogenetic and molecular studies have pointed to the possibility that at least some subsets of IMFT are, in fact, true neoplasms. However, one subset of IMFT is mostly probably associated with infection.7,8

Histologically, IMFTs are composed of myofibroblastic spindle cells that are mixed with a prominent infiltrate of lymphocytes, plasma cells, and acute inflammatory cells.1 The three basic histologic patterns were described by Coffin et al19 as follows: (1) myxoid, vascular, and inflammatory areas resembling nodular fascitis; (2) compact spindle cells with intermingled inflammatory cells resembling fibrous histiocytoma; and (3) dense platelike collagen resembling a desmoid or fibrous scar.

IMFTs of the larynx are very rare, with only 18 cases having so far been described in the literature.3 Reports of the imaging findings of IMFT are extremely rare.10 These tumors have primarily affected adults; they were localized lesion-producing regional but not systemic symptoms; they also had a predilection to the glottis and an indolent behavior.10 In our case, the patient had a long history of voice change and no systemic symptoms. Included in the major differential diagnosis is soft tissue tumor such as angioleiomyoma, leiomyoblastoma, or leiomyosarcoma and malignant tumor such as lymphoma, malignant stromal tumors (eg, malignant fibrous histiocytoma) or spindle cell squamous carcinoma.11,12 The current trend for management is laser excision with or without steroid therapy, and hemilaryngectomy is usually kept in reserve for multiple recurrences. Chemotherapy and radiation therapy have been reported for a few cases of recurrent IMFT or malignant transformation.9,13,14

Our case shows two interesting features. First, the signal intensity of the lesion was mixed hyper-intense and hypointense on T2-weighted images. However, a recent report of laryngeal IMFT by Munozo et al10 stated that the signal intensity of a subglottic lesion was isointense with the other soft tissues in all-conventional sequences. We assumed that the portion of the high signal intensity on T2-weighted images of our case may reflect the myxoid, vascular, or active inflammatory area, and the area of the low signal intensity on T2-weighted images may be a dense fibrous area. Second, the mean magnetization transfer ratio of the lesion was 0.347. The higher the MTR value of the lesion, the greater degree of suppression of the signal intensity after the MT pulse, and there is usually the more macromolecular cell wall protein in such a lesion.15 In our previous report, we suggested that a high MTR value for the lesion (>0.3) of the head and neck should be regarded as a malignant tumor, and this MTR finding generally has a high sensitivity (91%) and specificity (93%).16 Thus, a malignant submucosal tumor was the favored preoperative diagnosis. In our opinion, the reason why our case showed this high MTR value was that the tumor mainly displayed high cellularity, with such elements as dense collagenous stroma, spindle cells, and inflammatory cells on the pathologic examination.

**CONCLUSION**

We report here on the case of a 52-year-old female patient with laryngeal IMFT that clinically and radiologically mimicked a malignant submucosal tumor. The lesion was a well-enhanced elongated mass on the contrast-enhanced CT scan, it showed
mixed high and low signal intensity on T2-weighted images, it was a well-enhanced lesion on gadolinium-enhanced T1-weighted images and had a high MTR. Laryngeal IMFT is a benign and very rare lesion that can be treated by laser excision with or without steroid therapy, and hemilaryngectomy can generally be avoided. The diagnosis of IMFT should be considered for a patient with a solid submucosal laryngeal mass that especially shows good enhancement, mixed high and low signal intensity on T2-weighted images, and a high MTR.

REFERENCES


