

Case Report

Inflammatory myofibroblastic tumor resembling advanced carcinoma in larynx: a case report

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Abstract: Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm, especially occurs in the larynx, and is classified as a borderline neoplasm with uncertain behavior. This study aimed to present and discuss the case of a patient with a rare inflammatory myofibroblastic tumor (IMT) as an advanced carcinoma in larynx. Case report: A 63-year-old man with persistent hoarseness for past three months, was admitted to our ENT department. Computed tomography (CT) scans revealed an oval shaped mass centered in the right supraglottic larynx with destructing thyroid cartilage. The patient was diagnosed IMT by histology exam of biopsy specimens. We performed an extended partial laryngectomy to completely resect the tumor with a temporary tracheotomy. We further followed him for 73 months postoperatively and no recurrence was observed. Conclusion: Rare laryngeal IMT can progress to malignant tumor. Conservative excision with free margin is recommended.

Keywords: Inflammatory myofibroblastic tumor, inflammatory pseudotumor, laryngology, larynx

Introduction

Inflammatory myofibroblastic tumor is a rare mesenchymal neoplasm with unknown etiology, typically arising in lung, retroperitoneum, and extremities [1, 2]. Few additional cases (Table 1) have been reported in the larynx over the last decade [3-14]. Although it is generally benign, IMT often shows local destruction or infiltrative features that mimic neoplastic process. Then, IMT is considered to be a soft-tissue tumor with intermediate neoplasm potential [15, 16].

Materials and methods

A 63-year-old man was admitted to ENT department with a 3-month history of hoarseness in December 2010. He was a nonsmoker and had no history of any trauma or previous operation.

We performed flexible transnasal laryngoscopy and confirmed a mucosal protrusion of the right aryepiglottic fold with impaired laryngeal mobility ipsilaterally (Figure 1). Axial contrast-enhanced CT scans during admission showed a relatively well-enhancing, oval shaped mass

centered in the right supraglottic larynx while breaking through the thyroid cartilage plate (Figure 2). No lymph node enlargement was observed in the neck. We diagnosed the patient as an inflammatory myofibroblastic tumor (IMT) basing on the histology exam of his biopsy specimens.

Results

The patient was undergone an extended partial laryngectomy under general anesthesia and the airway was secured by a tracheotomy at the beginning of the procedure. The lesion was excised with free margin including the involved surrounding strap muscles.

Immunology and molecular biology findings

Grossly, the tumor was soft and with complete capsule, measuring in diameter 3.0 × 2.0 × 1.7 cm. A postoperative histopathological exam showed abundant spindle shaped myofibroblasts tightly arranged in fascicles, with scattered lymphocyte and plasma cells, and the right thyroid cartilage erosion (Figure 3). Cellular pleomorphism was demonstrated, and mitoses

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Table 1. Report cases of Laryngeal IMT in English literature in recent 10 years

No	Authors	Age (y)/sex	Location and size	Clinical information	Treatment and follow-up
1	Zitsch et al [3]	33/F	Left subglottic, 1.3 × 1.2 cm	Globus sensation; shortness of breath; stridor	Prednisolone × 6 wk followed by radiotherapy; open excision NED after 4 mo
2	Zitsch et al [3]	57/F	Left true vocal cord and subglottis	Progressive hoarseness for 1.5 y	Microaryngoscopic biopsy followed by CO ₂ laser ablation and prednisolone × 3 mo; NED after 3 mo
3	Idrees et al [4]	57/M	Anterior commissure 1.1 × 0.9 cm	Progressive hoarseness, 9 mo duration; history of traumatic intubation for a vocal cord cyst	Laryngoscopic excision with buccal mucosal graft; NED after 27 mo
4	Idrees et al [4]	28/M	Right vocal cord, 3 × 1.7 × 0.7 cm	Hoarseness 2 y, recently worsened	Complete surgical excision; NED after 18 mo of follow-up
5	Barreto et al [5]	22/M	Right vocal cord	Persistent hoarseness 2 mo	Laryngeal microsurgery; recurrence in 6 mo treated by laryngeal microsurgery; NED after 12 mo
6	Volker et al [6]	56/M	Left false cord, 2.7 cm	Dysphonia for 6 mo	Laser excision; No progression of the tumor after 24 mo
7	Volker et al [6]	34/F	Right vocal cord, 0.8 cm	Increasing dysphonia for 1 mo	Laryngeal microsurgery; recurrence in 3 mo treated by laryngeal microsurgery; NED after 31 mo
8	Alhumaid et al [7]	38/M	Subglottic area, 1.2 × 1.0 cm	Dyspnea and stridor for 3 y, recently worsened and hoarseness	Laryngeal microsurgery after tracheotomy followed oral corticosteroids and proton pump inhibitor therapy for 2 w; NED after 24 mo
9	Alhumaid et al [7]	54/M	Left vocal cord, 1.5 × 1.0 cm	Progressive hoarseness	Laser excision, recurrence in 12 mo treated by laser excision; NED after 6 mo
10	Dava et al [8]	73/M	Left vocal cord and subglottic area, 1.7 × 1.1 × 0.7 cm	Persistent hoarseness and progressive dyspnea for 1 y	Median vertical thyrotomy; NED after 8 mo
11	Do et al [9]	37//M	Right vocal fold	Hoarseness and dysphagia for 1 y	Laryngeal microsurgery followed oral steroids therapy for 4 w; NED after 6 mo
12	Kieu et al [10]	5/F	Subglottic area, 1.7 × 1.1 × 0.7 cm	Persistent episodes of cough and shortness of breath for 4 mo	Laser excision; NED after 12 mo
13	Tantau et al [11]	81/M	Right vocal fold	Hoarseness, dysphagia and fatigue for 4 y	Endoscopic excision; NED after 6 mo
14	Yan et al [12]	73/M	Right vocal cord	Hoarseness and cough for 2 mo	Laser excision; NED after 12 mo
15	Tay et al [13]	12/F	Right vocal cord, 1.5 × 1.6 × 1.7 cm	Hoarseness for 9 mo	laryngeal microsurgery; NED after 12 mo
16	Izadi et al [14]	46/F	Laryngeal surface of the epiglottis, 3.0 × 2.5 × 2.0 cm	Hoarseness for 4 y	Laser excisions twice before extended supraglottic laryngectomy; NED 12 mo after definitive surgery

NED indicates no evidence of disease.

were absent. The stromal cells were highly positive for vimentin, smooth muscle actin (**Figures 4, 5**) but have focally weaker positivity for anaplastic lymphoma kinase (ALK), muscle specific actin, S-100, Bcl-2, P53 and Ki-67 (**Figure 6**).

Follow-up

The patient was successfully decannulated 21 days postoperatively. Seventy-three months postoperatively, flexible transnasal laryngoscopy revealed no recurrence (**Figure 7**).

Discussion

IMT is rare and uncommon in head and neck especially in the larynx, although it has been

observed in the meninges, skull base, upper respiratory tract, orbits, tongue, ears, parapharyngeal space, and paranasal sinuses [16]. IMT is classified as a borderline neoplasm with uncertain behavior. Its etiology and pathogenesis remain unclear so far [17].

IMT can affect any subdivision of larynx, but the true vocal folds are the most common laryngeal site. Patients with laryngeal IMT generally present with hoarseness of varying severity and duration, or some nonspecific symptoms, including dysphonia, dyspnea, and stridor [3-9, 11-14]. Although the radiologic findings of IMT are nonspecific, particular findings are observed. IMT appears to be a well-enhanced elongated mass on the contrast-



Figure 1. Photograph showing mucosal protrusion of the right aryepiglottic fold.

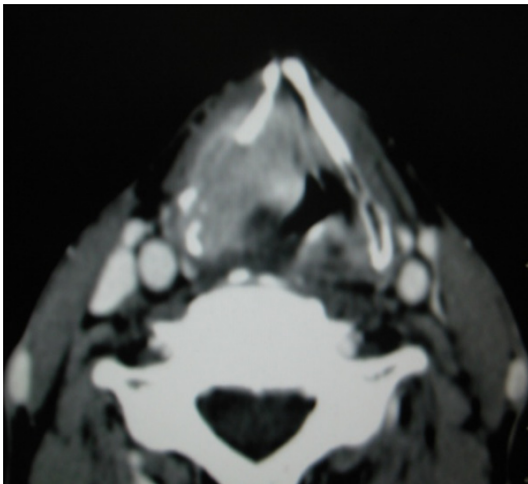


Figure 2. CT scan shows a relatively well-enhancing, oval shaped mass centers in the right supraglottic larynx with breaking through the thyroid cartilage plate.

enhanced CT scan. In uncommon cases IMT can invaded into surrounding muscles with osteolytic bone-destructive growth pattern [2, 16]. In this case, mimicking an advanced carcinoma (like cT4) seems to be very rare because most IMFTs are described as exophytic/polypoid lesions [4, 5, 7, 8, 12, 13].

Immunohistochemistry is usually utilized to confirm the myofibroblastic phenotype of the tumor spindle cells. The stromal cells in IMT are usually positive for muscle-specific actin, vimentin, and smooth muscle actin (SMA) [4,

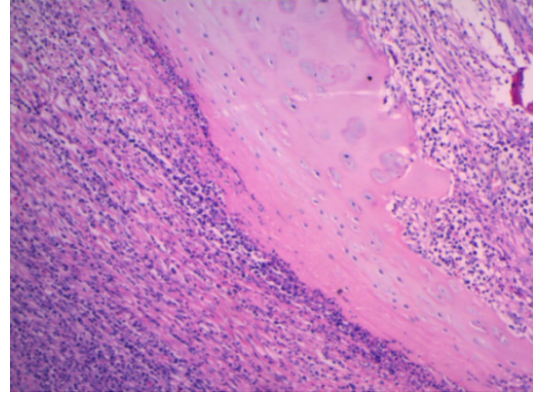


Figure 3. Postoperative histopathological exam shows abundant spindle shaped myofibroblasts tightly arranged in fascicles, with scattered lymphocyte and plasma cell, and the right thyroid cartilage erosion (H&E×100).

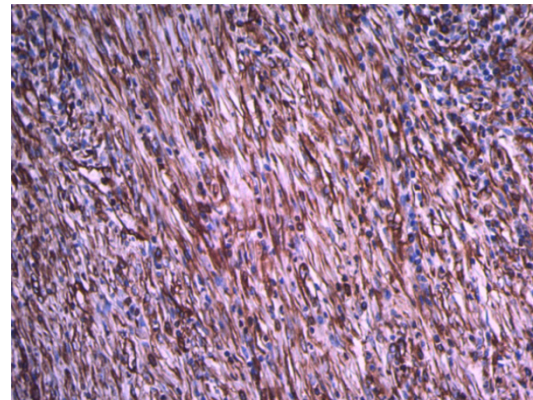


Figure 4. Immunohistochemically stromal cells shows a strongly positive reaction for vimentin (immunohistochemistry sp×100).

7-9, 13, 14]. Other myogenic markers positivity are less consistent including desmin, cytokeratin, and CD68. IMTs are typically negative to myoglobin and S-100 protein [4, 6-9, 12-14, 16]. In this case, the positive expression of P53, Bcl-2 and Ki-67 indicated the malignant tendency.

Most of the IMTs reported in head and neck region showed a benign course. However, cases related to paranasal sinuses seem to have highly aggressive behavior, with poor response to surgery, radiotherapy, and chemotherapy, also with multiple recurrences and fatal outcome [18]. For the laryngeal IMT, endoscopic excision with or without laser is considered to be the first line of treatment [3-7, 9-14]. If excision would result in significant function

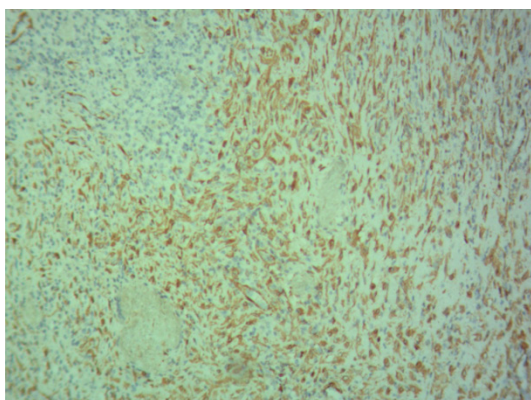


Figure 5. Immunohistochemical staining of the stromal cells with a strongly positive reaction for smooth muscle actin (immunohistochemistry sp×100).

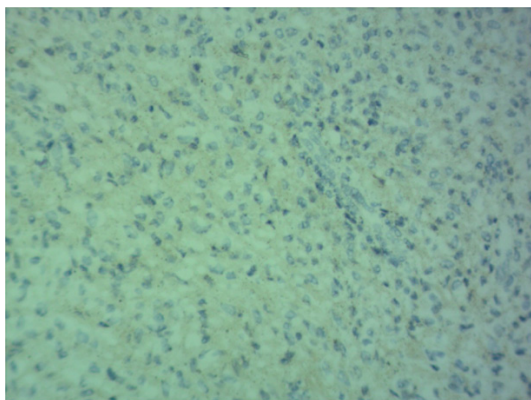


Figure 6. Immunohistochemically stromal cells shows a weaker positive reaction for anaplastic lymphoma kinase (ALK) (immunohistochemistry sp×100).

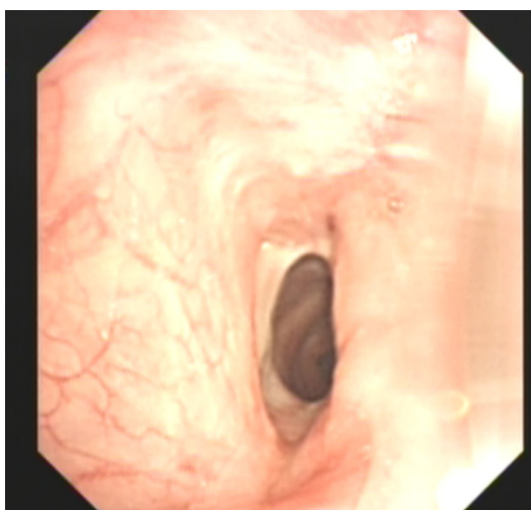


Figure 7. Seventy-three months postoperatively, flexible transnasal laryngoscopy reveals the right half laryngeal mucosa smooth and no recurrence had been noticed.

loss, a course of steroids alone may be attempted. Open excision should be reserved for cases of recurrence, poor endoscopic visualization prohibiting complete excision of the mass or when malignancy cannot be excluded [3]. Chemotherapy has been reported for a few cases of recurrent IPT or malignant transformation [4]. Steroid or radiation therapy as sole treatment seems less effective [3]. Focal gross positive margin can be treated by Gamma-Knife/CyberKnife Stereotactic Radio Surgery after surgical debulking [2]. In this case, due to the malignant tendency, surgical excision with free margins was the treatment of choice, and it preserved the laryngeal function simultaneously.

Some studies reported that IMT of larynx can be recurrent [5-7, 14]. The factors that can lead to recurrence may include partial excision, incomplete excision, or steroid therapy alone. Clinicians' strict follow-up is necessary, because there are no morphological features that can predict its biological behavior.

Conclusion

Rare laryngeal IMT can lead to malignant lesions. Histopathological and immuno-histochemical assessments are necessary for diagnosis. Conservative excision with free margin is recommended.

Disclosure of conflict of interest

None.

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