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*Case Report*

# Laryngeal adenoid cystic carcinoma: a rare case report

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**Abstract:** Laryngeal adenoid cystic carcinomas (LACC) are extremely rare and only account for less than 1% of all malignant laryngeal tumors. This tumor commonly occurs in the subglottic region of larynx, so dyspnea and hoarseness are its most common presenting symptoms. Adenoid cystic carcinoma is characterized by slow progression, late recurrence, and late distant metastasis. Total or partial laryngectomy is its treatment. Although it does not respond to radiotherapy completely, adjuvant radiotherapy or chemotherapy may be considered. In this study, we report a 41 year-old man, who had a rare recurrence of LACC, and we evaluate his clinical and pathologic characteristics.

**Keywords:** Adenoid cystic carcinoma, larynx, malignant tumor, case report

## Introduction

Laryngeal cancer is the second most common malignancy of the upper aero-digestive tract, with over 13,000 cases annually reported in the United States alone [1]. Although most (85-95%) malignant laryngeal tumors are squamous cell carcinoma (SCC), a wide variety of tumors of other histologic types may occur in the larynx [2].

Laryngeal adenoid cystic carcinomas (LACC) are rare and account for fewer than 1% of all malignant tumors occurring in the larynx. According to literature, fewer than 100 case reports are published on LACC worldwide [3].

The majority of these tumors occur in the subglottic (60%) and the supraglottic (35%) areas, and this may be due to the distribution density of sub-epithelial glands being higher in the supraglottic and subglottic areas compared to the glottis [4, 5].

Dyspnea and hoarseness are the most common presenting symptoms of LACC. Of note, the gender distribution of this tumor is equal [3, 4].

Histologically, ACC consists of cribriform, tubular, and solid subtypes. In all these three types, the cells are surrounded by hyaline or mucoid material, which gives it a cribriform or lacelike pattern [6].

LACC has an infiltrative growth pattern, a tendency for perineural invasion, and a susceptibility for hematogenous spread. Moreover, LACC tends to infiltrate into the surrounding structures in a slow manner. Furthermore, sometimes late recurrence or distant metastases can be found [6, 7].

The treatments proposed for LACC are controversial. In this regard, total laryngectomy is recommended by majority of otolaryngologists as the standard treatment for LACC, while partial laryngectomy can be performed occasionally. Although LACC is a relatively radioresistant tumor, chemotherapy, as an adjuvant or neoadjuvant therapy, may be useful for high-grade LACCs [8, 9].

The incidence of laryngeal adenoid cystic carcinoma (LACC) is reported as 0.005 per 100,000 population. In addition, few LACC patients have been confirmed in Iran in last decades [10].

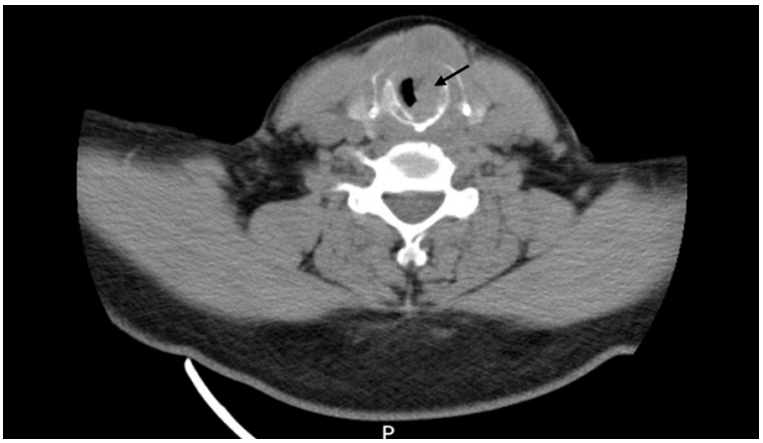

Figure 1. CT scan of larynx showing a subglottic submucosal mass that extended to the cricoid (arrow).

Because of the above-mentioned reasons and because the recurrence of LACC is rare, in the present study, we present the clinical and pathologic aspects of a rare case of laryngeal adenoid cystic carcinoma with recurrence, that was treated in our center.

## Case presentation

The patient was a 41-year-old man diagnosed with laryngeal adenoid cystic carcinoma (LACC) 7 years ago, which was treated with chemotherapy at that time. He referred to our center with a one-month history of dyspnea, hoarseness, respiratory distress, and stridor. There was no history of weight loss, odynophagia or dysphagia and nasal regurgitation. The patient's medical history was gastroesophageal reflux and he was a cigarette smoker.

Since we suspected recurrence of the previous cancer, the patient underwent a new investigation. On physical examination, the patient had no palpable neck mass or lymphadenopathy.

Indirect laryngoscopy detected a posterior left para-median sub-glottis submucosal mass. The laryngeal mucosa was found to be intact, and there was no necrosis in the laryngeal mucosa. Of note, both the glottis and supraglottic areas had a normal appearance. There was no impairment in laryngeal motion.

The patient underwent computerized tomography (CT) scan of the neck, which showed a submucosal mass in the left sub-glottic area that extended to cricoid and thyroid cartilage into the extra-laryngeal soft tissues on the left side of his neck (**Figure 1**).

Neither physical examination nor neck CT scan detected any evidence of node involvement in this patient. Chest X-ray findings were normal as well.

Next, we planned for a direct laryngoscopy evaluation and biopsy. On direct laryngoscopy, a mass was detected on the left posterior side of subglottis that extended to cricoid. Biopsy from the mass was sent for pathology. Histopathological examination reported the recurrence of adenoid cystic carcinoma, that showed three defined patterns: cribriform, solid, and less frequently tubular. The cribriform type was characterized by nests of cells with cylindromatous microcystic spaces. Moreover, the stroma was myxomatous and hyalinised, and perineural invasion was absent. To confirm this diagnosis, immunohistochemistry (IHC) was performed that was positive for p63 and cytokeratin (CK) 7, but negative for CK 20 (**Figure 2**).

Afterward, the patient underwent total laryngectomy and total thyroidectomy without neck dissection. Final pathology showed adenoid cystic carcinoma with surgical margins free of tumor, with no perineural invasion, but the left lobe of the thyroid was involved by tumor.

Seven days after operation, the patient was discharged. Thereafter, he was scheduled for adjuvant radiotherapy and during 2 years' follow-up, there was no evidence of any recurrence or metastasis.

## Discussion

Laryngeal adenoid cystic carcinomas (LACC) is rare, and constitutes less than 1% of laryngeal tumors. This tumor is characterized by slow progression, late recurrence, and distant metastasis [3, 7].

According to previous studies in this field, the most common primary site of LACC is the subglottic region (about 60%), followed by the supraglottic and the glottic areas, respectively.

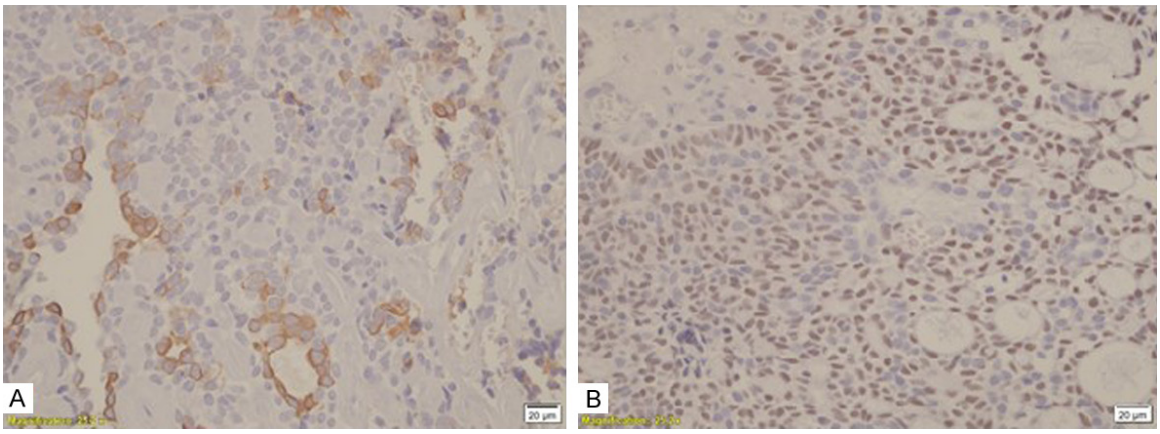


**Figure 2.** Immunohistochemical stains. (A) Cytokeratin 7 stained the luminal epithelial cells rimming the mucopolysaccharide filled spaces (×200), (B) p63 stained the nuclei of outer myoepithelial cells (×400).

Therefore, the signs and symptoms of LACC are related to anatomic location. Tumors involving the supra-glottis present with dysphagia. In addition, hoarseness is an indicator for glottic involvement. Stridor, dyspnea, and airway obstruction are more frequently associated with subglottic tumors [4, 11, 12]. In our case, the tumor was subglottic and presented with the common symptom of dyspnea.

Histopathological analysis is essential because the symptoms do not differ greatly from squamous cell carcinoma (SCC). Histopathologically, three patterns are seen in ACC: cribriform, tubular, and solid. Cribriform pattern is the most common, and has the best prognosis. The solid pattern is the least common one and has an aggressive course [13]. In our case, adenoid cystic carcinoma contained sheets of small uniform cells arranged in a classic cribriform growth pattern, which is the most frequent type.

In this case, first, we suspected a tumor based on the result of CT scan in left subglottic area (**Figure 1**), which was confirmed by the histopathologic evaluation.

The treatment options of laryngeal adenoid cystic carcinoma remain controversial. A total laryngectomy is the treatment of choice for LACC in the subglottic region, which was performed in our case. However, performing partial laryngectomy is also possible in selected patients with small, well-defined tumors and negative surgical resection margins or for LACC occurring in the supraglottic region. In addition, neck dissection should be performed in patients who are clinically or histologically confirmed to have nodal metastases [14-16]. Because LACC is often radioresistant, radiotherapy alone plays a minor role in treatment usually, but chemotherapy shows a positive response in advanced cases [8].

However, some studies have shown that combined treatment with conservative surgery and both post-operative radiotherapy and chemotherapy is necessary to reduce locoregional recurrence. Complete tumor removal is often difficult and local recurrences and distant metastases of tumor may be detected some years later. Thus, regular close and long-term follow-up is mandatory to detect late relapse and metastasis. In the present case, we observed a recurrence with thyroid involvement 7 years after chemotherapy. We performed total laryngectomy and thyroidectomy for this patient as well as radiotherapy after surgery and he was still alive without any distant metastasis after 2 years of follow-up.

## Conclusion

Adenoid cystic carcinoma of the larynx is a rare malignant neoplasm occurring in the subglottic area. Surgical resection with or without chemotherapy is also recommended. Late recurrence and distant metastasis are found in some cases. Therefore, long-term follow-up and surveillance should be done.

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## Disclosure of conflict of interest

None.

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