

# Adenoid cystic carcinoma in ventricle of larynx

## An interesting case

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### Abstract

**Rationale:** Adenoid cystic carcinomas (ACCs) are malignant tumors and occur in the major and the minor salivary glands. ACCs are rare in the larynx.

**Patient concerns:** A 55-year-old female patient who presented with a 12 months history of paraesthesia pharynges and hoarseness for 4 months. Laryngoscopy showed an endophytic lesion in the right supraglottic area with no laryngeal stenosis. And magnetic resonance imaging (MRI) confirmed the presence of a submucosal mass in the supraglottic area. Supporting laryngoscope was performed under general anesthesia and a lesion biopsy obtained.

**Diagnoses:** The lesion was sent for frozen-section biopsy, which shows the lesion was the epithelial-derived tumors of salivary gland, but it was impossible to identify the relationship between the tumor and the surrounding normal tissue, and to judge the nature of the tumor further. So, the lesion was sent for the second frozen-section biopsy, an ACC of the larynx was confirmed and margins were negative.

**Interventions:** The patient underwent partial laryngectomy.

**Outcomes:** The patient was doing well except hoarseness during the follow-up period with no evidence of disease recurrence or metastasis for 17 months post-operatively.

**Lessons:** Laryngeal ACC is a rare entity. When the nature of lesion cannot be confirmed, multiple biopsies may be required for confirm the diagnosis of pathology. It not only reduces patient's waiting time for surgery, but also define the diagnosis and surgical removal of lesion through a single anesthesia

**Abbreviations:** ACC = adenoid cystic carcinoma, MRI = magnetic resonance imaging.

**Keywords:** adenoid cystic carcinomas, larynx, surgery

## 1. Introduction

Adenoid cystic carcinomas (ACCs) are the most common malignant tumors that occur in minor salivary glands. A laryngeal location is rare because of the paucity of accessory salivary glands in this area. ACC account for less than 1% of all malignant tumors in the larynx.<sup>[1]</sup> Two-thirds of these laryngeal tumors occur in the subglottis; however, they also occur in the supraglottis in the false cords, aryepiglottic folds, and caudal aspect of the epiglottis.<sup>[2–3]</sup> The etiology of ACCs remains unknown. These tumors are found mainly in the fifth and sixth decades of life with a slight female predominance.

Two-thirds of these laryngeal ACCs occur in the subglottis. According to Dexamble et al, 64% of laryngeal ACC cases occur in the subglottis, 25% in the supraglottis, 5% in the glottis, and 6% in the transglottic area.<sup>[4]</sup> The histopathological pattern of ACC is classified into 3 distinct subtypes: cribriform, tubular, and solid.<sup>[5]</sup> This study aimed to present aspects concerning the clinical diagnosis and treatment with ACC, especially the diagnosis of frozen-section biopsy was ambiguous.

## 2. Case report

The case report was approved by the Ethics Committee of the First Affiliated Hospital, College of Medicine, Zhe Jiang University (approval no. 2017011). The written informed consent was obtained from the patient.

A 55-year-old woman who was a non-smoker was referred to our clinic with a 12-month history of paraesthesia pharynges and hoarseness for 4-month. The patient had no history of cough, prelaryngeal pain, dyspnea, or dysphagia. Laryngoscopy showed an endophytic lesion in the right supraglottic area with no laryngeal stenosis (Fig. 1). There was no impairment of the laryngeal motion. On physical examination, the patient had no palpable neck mass or lymphadenopathy. And magnetic resonance imaging (MRI) confirmed the presence of a submucosal mass in the right supraglottic area (Figs. 2 and 3). Neither physical examination nor neck MRI detected any evidence of node involvement. The findings from chest computed tomography (CT) were normal. It is difficult for biopsy under local anesthesia because the lesion lies in submucosal of the supraglottic area.

Editor: N/A.

The authors have no conflicts of interest to disclose.

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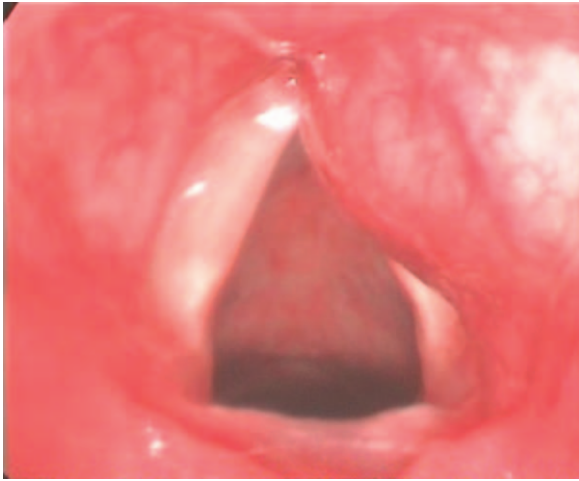
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Medicine (2018) 97:50(e13622)

Received: 23 August 2018 / Accepted: 19 November 2018

<http://dx.doi.org/10.1097/MD.0000000000013622>



**Figure 1.** Laryngoscopy image showing a endophytic lesion in the right supoglottic area with no laryngeal stenosis.

So, supporting laryngoscope was performed under general anesthesia and a lesion biopsy obtained. The lesion was sent for frozen-section biopsy, which shows the lesion was the epithelial-derived tumors of salivary gland, but it is impossible to identify the relationship between the tumor and the surrounding normal tissue, and to judge the nature of the tumor further. So, the lesion was sent for the second frozen-section biopsy, which shows the lesion was consistent with an ACC. Then, we performed partial laryngectomy without neck dissection. The surgical margins were free of tumor.

Specimen showed intra-luminal (sub-mucosal) growth without involvement of thyroid cartilage and muscular invasion (Fig. 4). Immunohistological staining showed that the tumor cells were positive for CD117, Calponin (CP), and P63 (Fig. 5). The final pathology report showed the specimen was consistent with an ACC of the larynx and negative margins.

Eight days post-operatively, the patient was discharged. And the patient was doing well except hoarseness during the

follow-up period with no evidence of disease recurrence or metastasis for 17 months post-operatively.

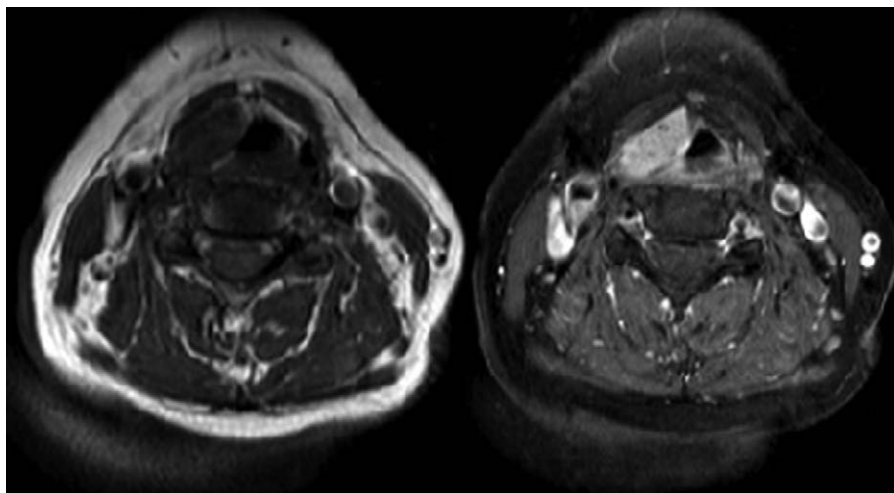
### 3. Discussion

ACC usually arise from subepithelial glands and most commonly present as submucosal masses. As a result, diagnosis is often delayed. ACC account for less than 1% of all malignant tumors in the larynx, and only about 120 cases have been reported in the literature until now.<sup>[1]</sup> These tumors have a slight female predisposition, and their peak incidence is in the fifth and sixth decades of life. The etiology of ACC remains unknown. There is no distinct risk factor that predisposes patients towards this malignancy. The signs and symptoms of laryngeal ACC are related to location and size.<sup>[6-8]</sup> Patients usually present with a history of mild to severe dyspnea at diagnosis. In the case, the patient was a 55 year-old woman who complain a 12-month history of paraesthesia pharynges and hoarseness for 4-month.

The histopathological pattern of ACC is classified into 3 distinct subtypes: cribriform, tubular, and solid.<sup>[5]</sup> When the nature of lesion cannot be confirmed, multiple biopsies may be required for confirming the diagnosis of pathology. In our case, 2 biopsies were performed, and the pathology was confirmed. It not only reduces patient's waiting time for surgery, but also defines the diagnosis and surgical removal of lesion through a single anesthesia.

Accurate pre-operative mapping and staging are essential. MRI is a very useful imaging procedure in ACC.<sup>[9]</sup> It is highly valuable in the assessment of primary tumor location, extra-luminal extension, regional, and distant metastases. It is, therefore, recommended in the pre-operative evaluation. MRI can underestimate the extent of the tumor because ACC may grow sub-mucosally without producing a distinct mass. In our case, MRI with contrast medium showed the presence of a sub-mucosal mass in the right supoglottic area without extension of thyroid cartilage and muscle. Also, these findings correlated with the histopathologic findings in this patient.

In view of the rarity of laryngeal ACC, the treatment options are still controversial.<sup>[10-12]</sup> Surgery with or without postoperative radiotherapy remains the mainstay treatment modality of



**Figure 2.** MRI scan of larynx showing supglottic submucosal mass and do not destruction at right side of thyroid cartilage. MRI T1-weighted hypointense that becomes hyperintensity on T1-weighted enhanced images. MRI=magnetic resonance imaging.

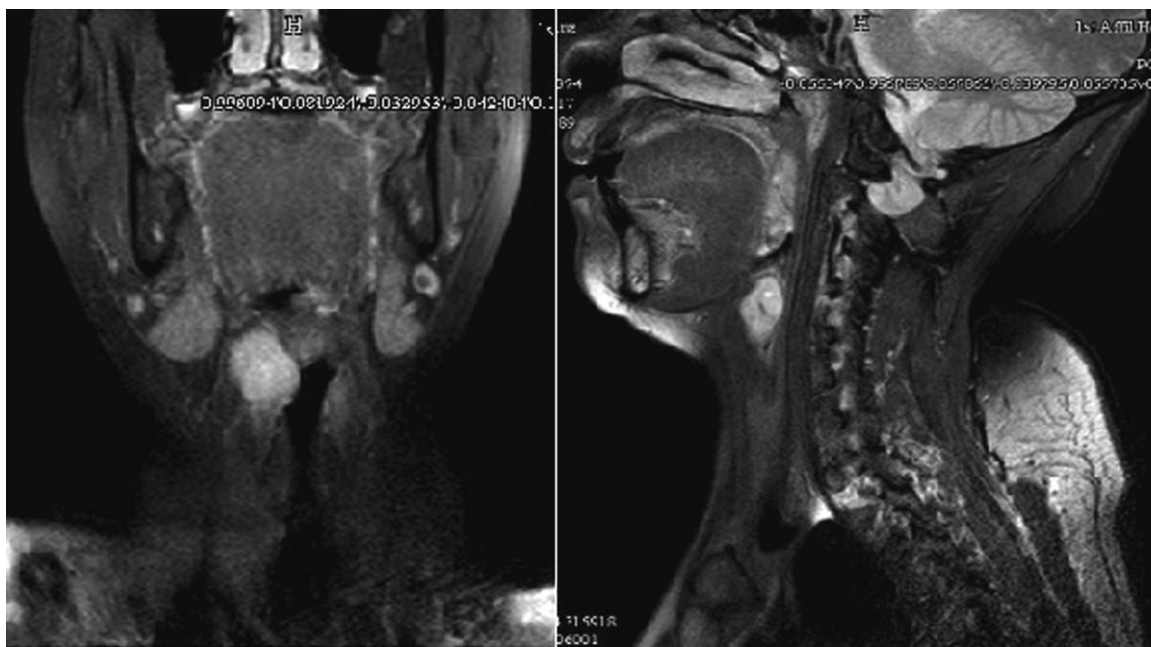


Figure 3. MRI T2-weighted scanning showed a high signal intensity mass in the right supraglottic area. MRI=magnetic resonance imaging.

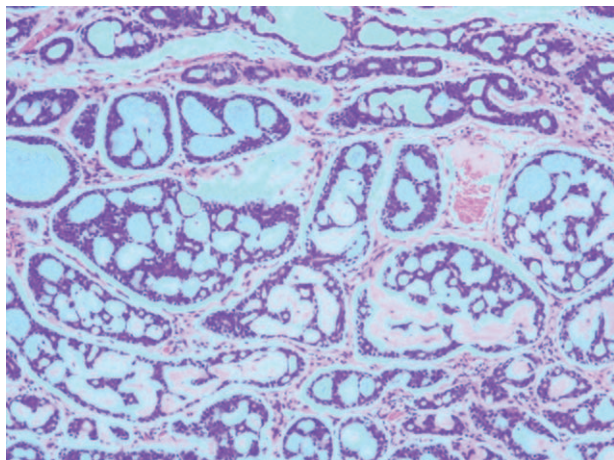


Figure 4. Photomicrograph of the tumor specimen showing the tumor cells were arranged in cribriform, mucous contained in glandular cavity, infiltrating growth, and tumor clinging to cartilage.(HE× 50).

laryngeal salivary gland carcinomas. Because of the risk of sub-mucosal spread and peri-neural and lympho-vascular invasion, total laryngectomy is often required and most authors agree that the treatment of choice is wide-margin local excision. Partial laryngectomy is possible in selected patients with small, well-defined tumors and negative surgical resection margins. In the absence of neck metastasis, elective neck dissection is not recommended. In our case, the pathology report showed negative margins, neither physical examination nor neck MRI detected any evidence of node involvement, so we performed partial laryngectomy without neck dissection.

Neck metastasis is rare, occurring in only 10% to 15% of the cases. Since early perineural and hematological spreading is common, local recurrences and distant metastases (especially to the lung) are common and sometimes arise years after the primary tumor has been diagnosed and treated.<sup>[13]</sup> Therefore, these patients require long-term follow-up. In the case, at 17-month's follow-up, there was no evidence of recurrence or metastasis.

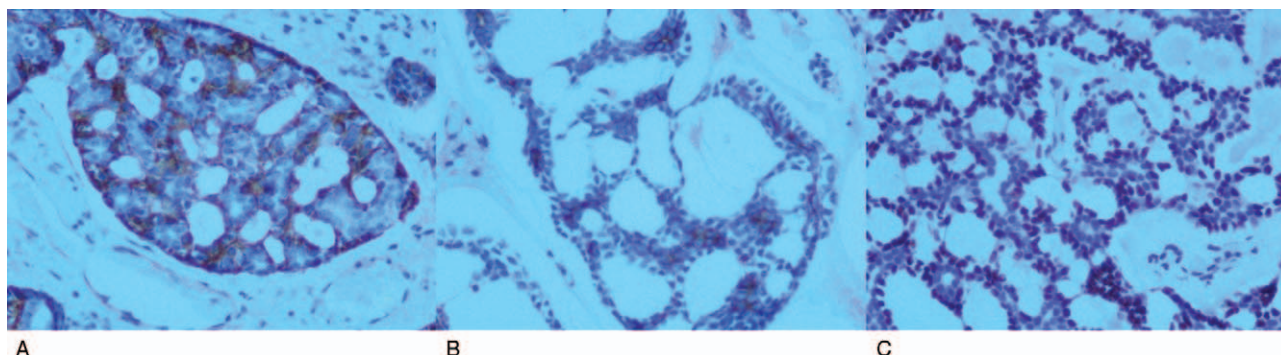


Figure 5. Immunohistological staining showed that the tumor cells were positive for Calponin (A), CD117 (B), and P63 (C).



#### 4. Conclusion

Laryngeal ACC is a rare entity and, tumor usually originates in the supraglottic or subglottic area with a predominance of old age women. MRI can be used to assess tumor extent and growth patterns. When the nature of lesion cannot be confirmed, multiple biopsies may be required for confirming the diagnosis of pathology. It not only reduces patient's waiting time for surgery but also define the diagnosis and surgical removal of lesion through a single anesthesia.

#### Author contributions

**Conceptualization:** wang qinying.

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**Methodology:** wang qinying.

**Resources:** wang qinying.

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**Writing – review & editing:** chen haihong.

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