Adenoid Cystic Carcinoma of the Larynx: A Common Tumor in a Rare Location

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Introduction

The salivary glands consist of major type, including the parotid, submandibular and sublingual glands, and minor type salivary glands. Minor salivary glands show the highest concentration in the hard and soft palate but are also found in the oral cavity, lips, tongue, oropharynx and larynx. Within the larynx the highest concentration of minor salivary glands is found on the false vocal cords and subepithelial surface of the anterior commissure. Malignancies of minor salivary glands can occur in any of the locations they are normally found. The most common histologic types of salivary gland malignancy are mucoepidermoid carcinoma, adenoid cystic carcinoma and adenocarcinoma with adenoid cystic carcinoma showing a frequency of 22% of all salivary gland malignancies. While adenoid cystic carcinoma is the most common salivary gland tumor to occur in the larynx this tumor is rare at 8% of all laryngeal malignancies. Within the larynx 6% of adenoid cystic carcinomas form in the subglottis, 25% are found in the supraglottis and the remaining 10% are glottic or tracheal. These tumors show equal prevalence among males and females and are most commonly found in the fifth to sixth decade of life. No distinct risk factors have been identified and unlike with other head and neck malignancies a correlation with tobacco use has not been observed. Presenting symptoms are dependent upon the location of the tumor with supraglottic lesions most commonly causing dysphagia, hoarseness or dyspnea is noted with glottic lesions and sublingual lesions causing stridor or airway obstruction. Close to one half of patients show vocal cord fixation on exam. 4. As the minor salivary glands of the larynx are subepithelial in location adenoid cystic carcinomas of the larynx presents grossly as a submucosal thickening often without an associated mass. The malignancy also spreads in this fashion without showing overt mucosal changes, resulting in the presentation of most patients at an advanced stage. When lymph node metastasis in the neck is rare, these lesions often show perineural invasion and the most frequent site of distant metastasis is the lung.5

Design

An unusual case of adenoid cystic carcinoma of the larynx with cervical metastasis is reviewed and presented.

Case Details

A 56-year-old woman presented with 6 months of shortness of breath and hoarseness associated with referred left-sided otalgia. She confirmed consumption of two to three alcoholic drinks per night and reported being an ex-smoker with cessation of use in 1978. Examination with flexible endoscopy revealed a subglottic mass with decreased mobility of the left hemilarynx. Dynamic videostroboscopy confirmed the mass involving the left supraglottic larynx with diffuse edema and submucosal thickening. The left vocal cord was paretic in the paramedian position and the vibratory wave was absent. The left hemilarynx demonstrated normal function and the vibratory wave was intact. MRI showed areas of nodular enhancement of the left vocal cord on T1 imaging measuring 7x7x9 cm with extension superficially surrounding the supraglottic area and base of the epiglottis as well as involving the left paratracheal sinus. This area was found to have an SUV of 4.68 on PET. Pathologic evaluation of biopsies of the left true and false vocal cords showed an infiltrative low-grade glandular neoplasm. This mass was favored to be a polymorphous low-grade adenocarcinoma of minor salivary gland origin notable for its rare location involving the larynx. Histologically, the neoplastic cells were uniform with oval nuclei and thin eosinophilic cytoplasm, surrounded by grayish blue hyalinized or myxoid stroma. The cells infiltrated the surrounding tissue in a single file fashion reminiscent of lobular carcinoma.

The patient underwent a total laryngectomy with bilateral neck dissections. Examination at that time confirmed mass effect of the malignancy at the level of the supraglottic larynx. Additionally, invasion into the pre-epiglottic space was noted and the subglottis was found to have an SUV of 4.68 on PET. Pathologic evaluation of the neck dissection specimens. Further observation regarding the propensity for nodal metastasis in low-grade versus high-grade laryngeal malignancies is necessary and could result in new recommendations regarding which patients would benefit from neck dissection at the time of resection.

Discussion

Adenoid cystic carcinoma most commonly shows an indolent clinical course with perineural spread notable for skip lesions of involvement. The majority of treatment is wide local excision at surgery with neck dissections for those with clinically, radiologically or histologically confirmed lymph node metastasis. In selected patients with high-risk attributes including positive surgical margins, perineural spread or high-grade malignancies adjuvant radiotherapy can be completed, as these lesions are radiosensitive although not usually cured by this modality alone. Improved outcomes have been noted with this multimodality treatment.3 For those found to have distant metastases at presentation combination chemotherapy with radiation therapy can be given for palliation.

In the literature regional lymphatic spread is discussed as being an uncommon occurrence with neck dissections not recommended for patients in an absence of established nodal spread. Our experience with the patient above differs from this more common experience as nodal spread was found on pathologic evaluation of the neck dissection specimens. Further observation regarding the propensity for nodal metastasis in low-grade versus high-grade laryngeal malignancies is necessary and could result in new recommendations regarding which patients would benefit from neck dissection at the time of resection.

Throughout the literature the reported overall five-year survival rate varies widely with survival prognosis differing by histologic subtype. The tubular subtype is considered to be the best prognosis, solid carries the worst and the cribriform or basaloid subtype, which is most common, is associated with an intermediate prognosis. Following treatment, long-term follow up is imperative as distant metastases to the lungs, liver, abdomen and bones are common and local recurrence can also occur with distant metastases showing the greatest impact on survival.5

Conclusion

Adenoid cystic carcinoma of the larynx is an indolent lesion, which grows in a submucosal fashion often without an associated mass and is a rare occurrence at 1% of laryngeal malignancies. As such, a high degree of suspicion must be maintained in hopes of detecting what is often already an advanced stage, most commonly of the supra- or sub-glottic larynx, at presentation. When appropriate radiologic and pathologic evaluations of the lesion have been completed the mainstay of treatment is wide local excision, most often by total laryngectomy, but also by partial laryngectomy as determined by the extent of the malignancy. At this time recommendations in the literature do not include elective neck dissection at the time of resection unless nodal enlargement is confirmed on examination or radiologic studies. However, our experience showed nodal involvement at the time of resection with completion of an elective neck dissection. This suggests that further investigation into the propensity for nodal spread based on the size or high or low histologic grading of the lesion could change this recommendation. Adjunct radiation therapy after resection is also used for lesions with high-risk characteristics. Local recurrences as well as distant metastases are common, for this reason long-term follow up is mandatory.

References