Adenoid Cystic Carcinoma of Larynx: Case Report

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Abstract

Introduction-Adenoid cystic carcinomas are malignant tumors that occur in both the major and the minor salivary glands. A laryngeal location is rare because of the paucity of accessary salivary glands in this area. Adenoid cystic carcinomas are rare account for less than 1% of all malignant tumors in the larynx, and only 120 cases have been reported in literature. **Case presentation-** We report a case of laryngeal adenoid cystic carcinoma in a 65- year-old female who presented with dyspnea, hoarseness and pain in throat. On examination growth over subglottis was present. Biopsy was taken and sends for Histopathological examination. The histopathological examination and IHC finding were suggestive of Adenoid cystic carcinoma of larynx. **Discussion-** Adenoid cystic carcinoma is very uncommon in larynx. These tumors have slight female predisposition and their peak incidence is in fifth and sixth decades of life. Features of diagnostic and therapeutic evaluation are described and clinical management of such cases is outlined.

Keywords: Adenoid cystic carcinoma, larynx, Salivary gland

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Conflict of interest-None

Introduction

Adenoid cystic carcinomas are malignant tumors that occur in both the major and the minor salivary glands. A laryngeal location is rare because of the paucity of accessory salivary glands in this area. Adenoid cystic carcinomas 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. These tumors have a slight female predisposition, and their peak incidence is in the fifth and sixth decades of life. There is no distinct risk factor that predisposes patients towards this malignancy. [1] The majority is subglottic (60%) or supraglottic (35%) and the true cords are involved in only 6% of cases. [2]

Case presentation

A 65- year- old, female was presented to our institution with 1 month history of dyspnea, hoarseness and pain in throat. On examination growth over sub-glottis and laryngeal strider present. Tracheostomy was performed to relieve dyspnea.

Neither physical examination nor ultrasonography of neck detected any cervical lymphadenopathy. The finding from chest X ray were normal

Direct laryngoscopy under general anesthesia showed endophytic growth present over posterior part of subglottis on left side and over inferior surface of left vocal cord which was bulge on the retroarytenoid region on left side. Left vocal cord was fixed and right vocal cord was mobile. Rests of the structures were normal. Biopsy was taken and sends for histopathological examination.

Gross examination-

We received multiple soft tissue bits labeled as a biopsy from subglottic growth collectively measuring 0.5 X 0.3 X0.2 cm3 in size, greyish in color.

On Microscopic examination-

The section showed hyper plastic squamous and columnar epithelium. Submucosa showed tumor cells arranged in tubular, trabecular and cribriform pattern with pseudocyst formation. Most of the cells were round with hyperchromatic, angulated nucleus and scanty cytoplasm. Few cells were round to polygonal with vesicular nuclei along with variable amount of clear cytoplasm. Stroma was hyalinized containing focal mucoid material. There was no evidence of perineural invasion in received biopsy bits.

IHC finding-

Tumor cells (epithelial cells) were immunopositive with EMA and CD117 (focal positive) and myoepithelial cells were

highlighted with S100, SMA (focal positivity)

Final diagnosis of Adenoid cystic carcinoma was given. The patient was referred to nearby cancer institute and was then lost to further follow up.

Discussion

Adenoid cystic carcinomas arise from subepithelial glands and most commonly present as submucosal mass. Because they spread in a submucosal fashion, they are difficult to detect earlier. This explains why most patients were diagnosed late, at the advanced stage. They tend to spread by perineural extension and frequently recur after initial treatment. Neck metastasis is rare. [3] The tumors are often painful, possibly because they frequently show perineural invasion. Adenoid cystic carcinoma often metastasizes to distant locations, especially the lung, unlike other salivary gland malignancies. [4]

The histological pattern of adenoid cystic carcinoma is classified into three subtypes: tubular, cribriform and solid. In the tubular form, well-formed ducts and tubules with central lumina are lined by inner epithelial and outer myoepithelial cells. The cribriform pattern, the most frequent, is characterized by nests of cells with cylindromatous microcystic spaces. These are filled with hyaline or basophilic mucoid material. The solid or basaloid type is formed of sheets of uniform basaloid cells lacking tubular or microcystic formation. Tumors consist of two main cell types: ductal and modified myoepithelial cells that typically have hyperchromatic, angular nuclei and frequently clear cytoplasm. Generally tumors composed of tubular and cribriform pattern pursue a less aggressive course than those with greater than 30% of solid component. [2]

Basaloid squamous cell carcinoma, polymorphous low-grade adenocarcinoma, pleomorphic adenoma, epithelial-myoepithelial carcinoma, basal cell adenoma or adenocarcinoma is often considered in differential diagnosis. On biopsy, most important distinction is versus basaloid squamous cell carcinoma. This is because of preoperative radiation and neck

dissection may be used with basaloid squamous cell carcinoma but should not be used with adenoid cystic carcinoma.

Histologically adenoid cystic carcinomas show less cytological pleomorphism than basaloid squamous cell carcinoma and fewer mitotic figures and should not contain areas of squamous differentiation or dysplasia of surface epithelium. The presence of nodal metastases virtually excludes the diagnosis of adenoid cystic carcinoma, as this tumor rarely metastasizes to lymph nodes.

Polymorphous low grade adenocarcinomas usually show more heterogenecity in their growth pattern and cytologic features, while adenoid cystic carcinoma tends to have smaller and more angulated nuclei. Adenoid cystic carcinomas have been shown to be much more likely stain with antibodies to c- kit. [4] Epithelial membrane antigen also be useful in separating PLGA from ACC. With this stain more than 90% of the tumor cells in PLGA are EMA positive, whereas in ACC, EMA stains only those cells lining lamina and not the non-luminal tumor cells or pseudocyst lining cells (IHC).[5]

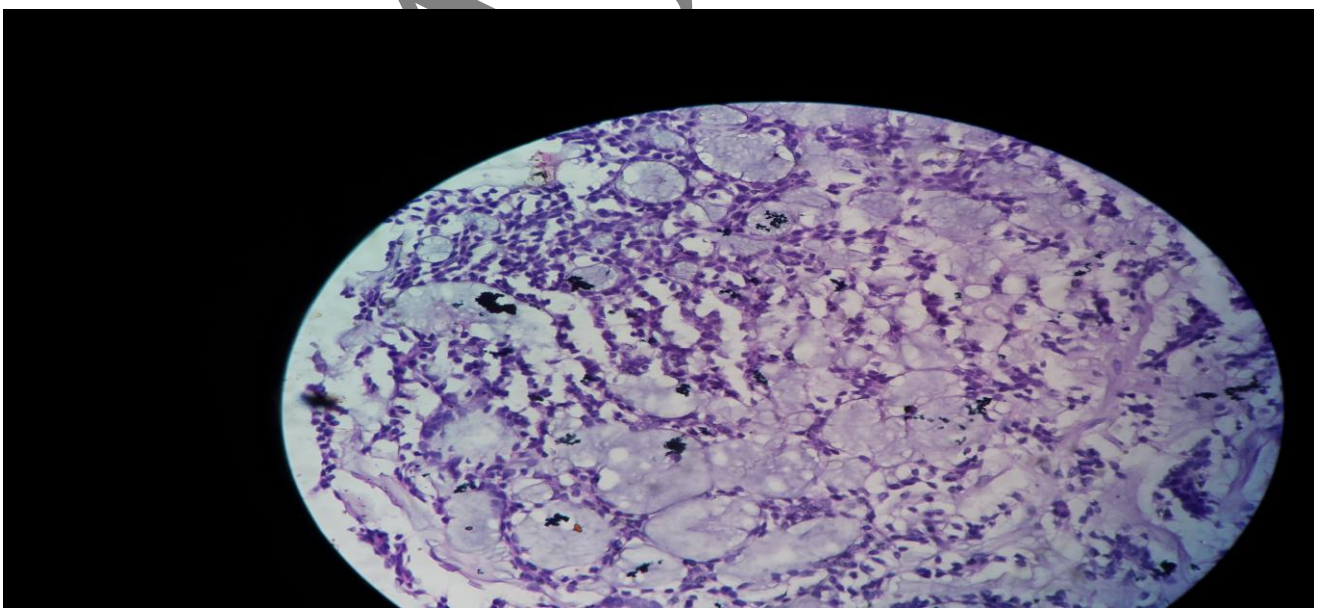

Figure: Tumor cells arranged in Cribriform pattern with Pseudocyst formation

In case of pleomorphic adenoma the identification of a mesenchymal component would be indicative of a pleomorphic adenoma, whereas identification of a perineural invasion or infiltration of surrounding tissues would be indicative of adenoid cystic carcinoma. [4]

In case of basal cell adenoma the characteristic cribriform pattern of adenoid cystic carcinoma is rare and only minor component of basal cell adenoma. Basal cell adenoma lack cells with irregular, angular shaped nuclei that are common and characteristic in adenoid cystic carcinoma. Infiltration and perineural invasion definitely distinguish adenoid cystic carcinoma from basal cell adenoma.

The cells in the adenoid cystic carcinomas are usually smaller with more irregular, angular, hyper-chromatic nuclei and characteristic cribriform pattern is not a feature of epithelial myoepithelial carcinoma. [6]

In view of the rarity of laryngeal adenoid cystic carcinoma, the treatment options are still controversial. The treatment of choice is wide-margin local excision (partial or total laryngectomy, depending on the location and size of the tumor). Radical neck dissection is indicated for patients who have clinically or histologically confirmed nodal metastases. These tumors have been shown to be radiosensitive but usually not radiocurable. Therefore, radiotherapy alone usually has little role in treatment. [1]

Conclusion: Adenoid cystic carcinoma of larynx is rare tumor and account for < 1% of laryngeal malignancies. Therefore, a high degree of suspicion is essential for early diagnosis. The correct recognition of this entity and differentiating it from other mimics are crucial to proper patient management and appropriate follow up. For these patients, regular and long-term follow-up is mandatory, in order to detect relapses and metastases.

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