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Case Report

Adenoid Cystic Carcinoma of the Trachea

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Abstract: Adenoid cystic carcinoma is a malignant tumor of salivary glands. It is extremely rare in trachea worldwide. They arise in the subepithelioal minor salivary glands, grow slowly and metastasize by perineural invasion. Patient usually presents with dyspnea. It is diagnosed by histopathological evaluation of biopsy taken by endoscopy. Its treatment includes surgical resection with or without radiotherapy. They frequently recur after initial treatment.

Keywords: Tracheal lesion, Adenoid Cystic Carcinoma, Minor Salivary Glands

Introduction

Tracheal adenoid cystic carcinoma (ACC) is exceedingly rare, representing 1% of all respiratory tract cancers. It is a slow growing tumor with a prolonged clinical course. The main presenting symptom is dyspnea therefore it is often misdiagnosed as bronchial asthma or chronic bronchitis. Treatment is surgical resection combined with radiotherapy because of close surgical margins. In inoperable cases radiotherapy results in long periods of remission [1]. We report a case of primary ACC of trachea of a 40 year old female who presented with a history of dyspnea for one year and was finally diagnosed as a case of adenoid cystic carcinoma on histopathological evaluation of bronchoscopic biopsy.

Case Report

A 40 years old female presented with one year history of progressive shortness of breath which worsened on lying down. She received treatment for asthma at local hospital but it did not relieve her symptoms. Her condition deteriorated and she presented to PIMS, main ER from where she was admitted in pulmonology ward. According to her medical history she was a non smoker with insignificant past medical or surgical history. On physical examination she had increased respiratory rate. She was mildly anaemic with oral thrush. There were no palpable lymph nodes. She was afebrile with normal pulse and blood pressure. On chest auscultation there was ronchi all over the chest.

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Her computed tomography revealed a tracheal mass with associated pneumomediastinum. Bronchoscopy was done which revealed a large lobulated mass in the middle trachea obstructing 90 % of its lumen. Multiple biopsies were taken and sent for histopathological examination.

Microscopic examination revealed a malignant neoplasm consistent with adenoid cystic carcinoma. The neoplasm is arranged in a cribriform pattern. The individual neoplastic cells were small bland myoepithelial cells with scant cytoplasm and dark compact angular nuclei surrounding pseudoglandular spaces. A diagnosis of adenoid cystic carcinoma was made.

Surgery has been done by end to end anastomosis.

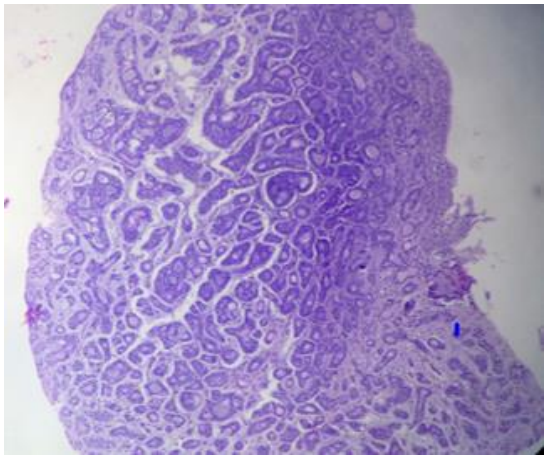

Figure-1: Low power view of one of the four fragments of the tumor showing cribriform pattern, typical of adenoid cystic carcinoma (H&E x 100)

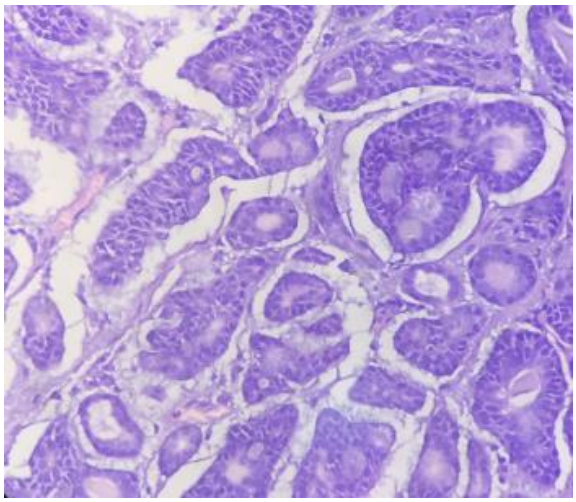

Figure-2: High power view of the tumor showing small bland myoepithelial cells with scant cytoplasm and dark compact angular nuclei surrounding pseudoglandular spaces. No nerve was seen in the available biopsy material (H&E x 400).

Discussion

Primary tracheal tumors are rare with incidence less than 0.2 per 100,000 persons per year, 10% of which are ACC(1). Kumar N, et al. reported six patients in India (2), El Marjany M, et al. reported two cases in Morocco (1), Yang PY, et al. reported seven cases in Taiwan (3) of adenoid cystic carcinoma of trachea. ACC was previously called cylindroma or adenocystic carcinoma. It occurs in young age, equally in both sexes and is unrelated to smoking. Most patients with tracheal ACC present with dyspnea, and the symptoms mimic those of bronchial asthma or chronic bronchitis (1).Tumors may not cause symptoms until 75% of the tracheal luminal diameter is occluded by the tumor (4)The main symptoms include hemoptysis, nonproductive cough, dyspnea, chest pain and weight loss (3).Diagnosis is made by histopathological evaluation of biopsy. Bronchoscopy is the standard diagnostic procedure for obtaining biopsy but in some selected cases Chamberlains procedure is also done[5].Computed tomography or magnetic resonance imaging typically shows longitudinal thickening of the tracheal wall extending superiorly and inferiorly from the main mass(6). Majority of the primary tracheal tumors arise in the distal third region of the trachea (7). This tumor characteristically grows into the airway lumen, forming a smooth surfaced polypoid growth tumor. Sometimes the growth is annular. Submucosal extension, sometimes to a considerable distance from main tumor can be found (8).

Malignant tumors of trachea are more common than benign tumors. Squamous cell carcinoma and Adenoid cystic carcinoma (ACC) are the most frequent histologic types, accounting for approximately two-thirds of primary neoplasms of the trachea. The tumor is slow growing but with distant metastasis [1].

Tracheal submucosa contains evenly distributed minor salivary glands which morphologically resembles minor salivary glands of oropharynx and develop pathologies similar to them but at a lower frequency [9]. ACC is the most common malignant neoplasm of the minor salivary gland followed by mucoepidermoid carcinoma, polymorphous low-grade adenocarcinoma and clear cell carcinoma [10].

The etiology of adenoid cystic carcinomas is unknown. The patients are diagnosed at late stages because they arise in sub-epithelial glands. They metastasize by perineural invasion and usually metastasize to lung [11].

ACC has three histopathological patterns; cribriform, the most common pattern; tubular, with the best prognosis; and solid, with worst prognosis [12].

The differentials of ACC include pleomorphic adenoma and low grade polymorphous adenocarcinoma. Histologically the pleomorphic adenoma has squamous metaplasia and mesenchyme like areas with no perineural invasion and polymorphous adenocarcinoma has bland uniform cells. Immunohistochemically, ACC is negative for GFAP and CD 57 while pleomorphic adenoma is positive for them and ACC is positive for CD 117,while its differential low grade polymorphous adencarcinoma will be negative for CD 117 [13,14].

The grading of the tumor is based on the presence or absence of solid component which determines its prognosis[15].

ACC is a non-encapsulated malignant tumor which metastasizes by direct extension and perineural invasion. It mostly metastasize to the lungs and less commonly to the brain, bone, liver, kidney, skin, abdomen; and heart.

Management include surgery and radiotherapy, alone; or in a combination. The surgery includes resection of tumor with end to end anastomosis. Because of local metastasis and presence of adjacent vital organs usually complete resection is not possible for which radiotherapy is done after surgery. Chemotherapy has no role in the treatment of ACC of the trachea.

Post surgical survival time range from 7.5years to 118 months [1].The 5- and 10-year survival rates of all types of tracheal cancer are 5%–15% and 6%–7% respectively, while those of patients treated with surgical resection are up to 50% and 35%–50% respectively emphasizing that diagnosis followed by treatment can enhance life expectancy as mentioned [16]..

Conclusion

ACC is a rare primary tracheal malignant tumor. The diagnosis is usually delayed because of misleading symptoms of bronchial asthma. Surgery followed by radiotherapy is the recommended treatment providing pronlonged survival [1].

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A. Conception/Study Designing/Planning
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D. Manuscript Writing
E. Critical Review
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