An Uncommon Primary Tracheal Tumor Masquerading as Chronic Obstructive Pulmonary Disease: Diagnosed at Autopsy

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Primary tracheal tumors are rare neoplasms and often are hard to diagnose early due to the non-specific clinical presentations. Prolonged symptomatic treatment for these can often lead to missing out the diagnosis and can prove fatal. A 35-year-old male presented with gradually increasing breathlessness and dysphagia to solids and was being treated with bronchodilators and antibiotics. Chest computed tomography (CT) revealed an intratracheal mass in the lower part of the trachea and the patient was referred to our institution in view of a primary tracheal tumor where he expired within 10 min of admission. On autopsy, the gross findings and histological examination revealed a diagnosis of adenoid cystic carcinoma in the trachea. To conclude, primary tracheal tumors, even though rare, should be suspected in cases of refractory chronic obstructive pulmonary disease and chest CT and bronchoscopy must be done to aid correct diagnosis.

Keywords: Adenoid cystic carcinoma, Chronic obstructive pulmonary disease, Intratracheal mass, Primary tracheal tumor

INTRODUCTION

Primary malignant tumors of the trachea are rare, accounting for only 0.1% of all malignancies.¹ The most common histological types of a primary tracheal tumor are squamous cell carcinoma followed by adenoid cystic carcinoma (ACC).² ACC referred to in older textbooks as cylindroma is a rare type of lung cancer arising from mixed seromucinous glands seen in tracheobronchial submucosa. It accounts for 20-25% of all tracheal tumors and 80% of all tracheobronchial gland tumors.³

We report a rare case that presented with dyspnea and dysphagia and was initially treated with bronchodilators, but was found to be ACC of the trachea on autopsy.

CASE REPORT

A 35-year-old male patient presented with gradually progressing dyspnea since the last 6 months and progressive dysphagia to solids since 1 month. He was being treated as a case of chronic obstructive pulmonary disease (COPD) with bronchodilators and antibiotics. Since there was no decrease in symptoms, a high resolution chest computed tomography (CT) was done, which showed an intratracheal mass 2 inches above the carina measuring 2 cm × 2 cm almost occluding the entire lumen (Figure 1).

The patient was referred to our institution where he expired within 10 min of admission and an autopsy was requested.

On autopsy, the trachea showed an intraluminal dumbbell shaped polypoidal mass measuring 2.5 cm × 2 cm, whitish and glistening. On cut surface, it was firm to cut and whitish in color with no areas of necrosis or hemorrhage (Figures 2 and 3).

The mass appeared to be arising from the lateral wall of the trachea. There was no other significant finding in any other organ except bilateral pulmonary edema. On histopathology, section from the tumor shows normal tracheal mucosa, beneath which were present infiltrating cribriform islands of tumor cells with small basophilic nuclei and surrounded by varying amount of eosinophilic and hyalinized stroma (Figures 4 and 5). The histomorphological features were consistent with primary ACC of the trachea.

DISCUSSION

Tracheobronchial tumors can be categorized as malignant or benign. Malignant can be either primary or secondary.⁴

Primary malignant neoplasms of the trachea are uncommon, accounting for <1% of all thoracic malignancies. The presentation is variable, and the initial symptoms are non-specific including dyspnea, wheezing, stridor, hemoptysis...
or dysphagia. These may often lead to a misdiagnosis of COPD and may be treated accordingly. The lesions may be missed on chest radiographs and are rarely identified prospectively.

The most common primary tracheal malignancies are squamous cell carcinoma and ACC. Less common tumors include mucoepidermoid carcinoma and carcinoid tumors.2

ACC, formerly known as cylindroma, was first described by Billroth in 1859 apart from dyspnea, stridor, and dysphagia, chest pain has been reported in the literature as a presenting symptom of tracheal ACC.5

Tracheal ACC demonstrates no racial or gender predilection and no clear association with smoking has been established.
The location of the tumor can be in any region of the trachea and may involve long segments at presentation.

Grossly the tumor is poorly circumscribed, white gray solid mass with infiltrating borders. Microscopically they have a characteristic cribriform pattern consisting of cytologically bland tumor cells arranged in cords or nests. True glandular spaces and pseudo glandular spaces filled with eosinophilic homogenous material are found within the cribriform structures. Perineural invasion is another characteristic feature.

At CT, ACC generally manifests as an intraluminal mass with soft tissue attenuation. Primary circumferential resection with end to end anastomosis stays the mainstay in therapy in cases of localized disease. Adjuvant radiotherapy is also recommended. In inoperable cases, palliative radiotherapy can be used.

Metastasis generally involves the lung, but may spread to extrathoracic sites as well.

**CONCLUSION**

Primary tracheal tumors, even though rare, should be suspected in cases of refractory COPD. In all cases of prolonged breathlessness and cough, chest CT and bronchoscopy must be done to aid correct diagnosis.

Primary tracheal tumors if diagnosed early, can be curable by surgical excision and/or chemotherapy based on the histological type and grade. Hence, a high index of suspicion and prompt investigation with appropriate management is required.

**REFERENCES**