

Adenoid Cystic Carcinoma Presenting as a Tracheal Polyp

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1. Abstract

Tumours primarily involving the trachea are very rare, Adenoid Cystic Carcinoma accounts for most cases after Squamous Cell Carcinoma. Adenoid cystic carcinoma of the airway originates from the submucosal glands of the tracheobronchial tree. These tumours pose a diagnostic and therapeutic challenge because of their slow growth and tendency for local recurrence and late metastasis. We present a case of adenoid cystic carcinoma of the trachea masquerading as a tracheal polyp clinically.

2. Introduction

Adenoid cystic carcinoma (ACC) also known as cylindroma is a malignant tumour of salivary gland origin, first reported by Billroth in 1856. These neoplasms most commonly involve the parotid gland and account for 10% of tumours of the head and neck. These tumours arise from both major and minor salivary glands; therefore can occur in any head and neck site containing salivary gland tissue. Primary tracheal tumours are extremely rare, accounting for approximately 0.2 % of the entire respiratory system and 80 % of these are malignant. Adenoid cystic carcinoma (ACC) is the second most common primary malignant tumour of the trachea, after squamous cell carcinoma. The most common signs and symptoms are those of upper airway obstruction including dyspnoea, cough, haemoptysis, and stridor which are often insidious, delaying diagnosis and optimal management [1-3].

3. Case Report

A forty-six-year-old female presented to the outpatient department

with complaints of coughing and wheezing for few months. The patient gave no history of fever or chest pain or any other history significant for suggesting laryngeal involvement. Her vital signs were within normal limits with no cervical or generalized lymphadenopathy. She was clinically euthyroid. There was no significant past or family history.

A pulmonary function test (PFT) done outside showed mild restrictive pattern and the treadmill test (TMT) result was negative. She had taken symptomatic treatment from local alternative medicine practitioners and was also started on anti-histaminics with no relief. Cardiovascular and respiratory systems also were clinically normal. There was no thyroid gland enlargement or thyroid cartilage tenderness, and no cricothyroid escape. Laryngeal crepitus was present. Upon performing laryngoscopy, a growth was observed over the anterior wall of trachea. An incisional biopsy was performed which showed a sub epithelial tumour composed of uniform bland basaloid cells arranged in a cribriform pattern with myxohyaline material & foci of hemorrhage & necrosis with histiocytic & giant cell reaction. The biopsy was reported as suggestive of a Salivary Gland Type Tumour – favor Adenoid Cystic Carcinoma (ACC), Cribriform pattern (Figure 1A, B). The patient subsequently underwent resection of the tumour. We received a segment of trachea measuring 4.0cm in greatest dimension. The luminal surface showed a polypoid ulcerated mass measuring 1.5 cm in greatest dimension. The histopathologic sections showed features of Adenoid Cystic Carcinoma, Cribriform type. Solid component comprised less than 5% of the total tumour. The dis-

tance from the closest superior resected margin was 06 mm. The tumour was limited to the trachea and the surrounding cartilages were uninvolved by the tumour (Fig 2A, B, C). The patient is on regular follow up for the last one year and is disease free till date.

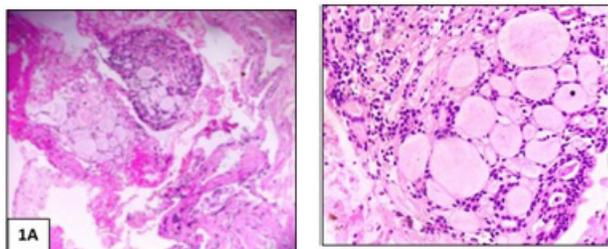


Figure 1: Incisional biopsy - Photomicrograph showing tumour composed of tumour cells arranged in cribriform pattern

- A) HE- 100x
B) HE- 400x

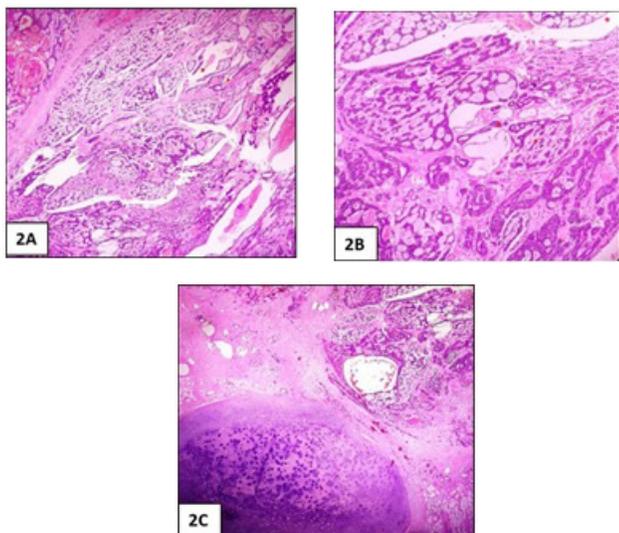


Figure 2: Resection specimen

- A) Photomicrograph showing tumour composed of tumour cells arranged in cribriform pattern. (HE-100x)
B) Photomicrograph showing tumour composed of tumour cells arranged in cribriform pattern with central myxohyaline material. (HE-100x)
C) Photomicrograph showing tumour cells close to cartilage but involving the tracheal cartilage. (HE-100x)

4. Discussion

Adenoid cystic carcinoma is a common salivary gland malignancy in the head and neck but rarely arises in the major airways. ACC of the larynx and trachea arise from the mixed seromucinous glands. These glands decrease from the supraglottis to the glottis, subglottis, and trachea. The false vocal cords and the area just distal to the anterior commissure have been shown to have the most glands. Despite these findings, ACC has been more commonly reported in the trachea than in other airway subsites [2, 3]. Clinical symptoms such as coughing, dyspnea and stridor are non-specific and

insidious and these tumours are often slow-growing. In the present case also the patient presented with similar complaints. Tracheal ACC is equally prevalent in males and females. Unlike SCC, no definite association with cigarette smoking is observed in ACC. The tumour is most common in patients in the 4th and 5th decades of life. Our patient was also a female in the 5th decade of life with no history of cigarette smoking. ACCs of the airway can be subcategorised as laryngeal or tracheal in origin (Calzada et al). The laryngeal subsite comprises the area between the epiglottis superiorly and the cricoid cartilage inferiorly. The tracheal subsite is defined as the area distal to the cricoid cartilage.

Grossly, these tumours characteristically grow into the airway lumen, forming a smooth surfaced, somewhat polypoid masses. Occasionally, the tumour is circumferential and annular. Submucosal extension, sometimes to a considerable distance from main tumour is not uncommon. Histologically, three patterns are seen; trabecular, cribriform and solid type. The cribriform pattern is most common consisting of uniform cells with relatively little cytoplasm arranged in well-defined nests of variable size. The cells in these nests are separated by well-defined cystic spaces containing a mucinous substance that stains strongly with alcian blue and weakly with Periodic Acid–Schiff (PAS).

ACC is a neoplasm with a prolonged clinical course and late onset of metastases and local recurrence. ACC spreads most commonly by direct extension, through submucosal or perineural invasion or hematogenous metastasis. More than 50% of patients with tracheal ACC have hematogenous metastases. It is also known for its neurotropic tendency, accounting for loco-regional recurrences many years after initial presentation and treatment [5, 6]. Pulmonary metastases are the most common distant metastases and may occur many years after treatment. However, occasional metastases have been reported to organs like liver, brain, bony skeleton, etc. Lymphatic spread of tracheal ACC is relatively uncommon. Lymph node metastases are reported in 10% of patients. 2,7-8 Our patient did not have evidence of metastases at the time of diagnosis and 12 months after follow-up.

Treatment options include surgery alone, radiation therapy alone, or a combination thereof. The surgical methods are primary tracheal resection and reconstruction, primary tumour resection, and endoscopic resection, either by coring or using a laser. Radiation therapy for ACC of salivary glands has been found to improve local control of tumours but does not affect survival. The efficacy of radiation has not been investigated for tracheal ACC. The role of post-operative adjuvant radiotherapy remains uncertain. It is reasonable to assume that adjuvant radiation therapy may be beneficial and is likely to delay or reduce the incidence of local recurrence in the airway if surgical removal is complete. According to the results of previous studies the 5-year survival ranges from 66% to 100% and the 10-year survival ranges from 51% to 62% for

patients with tracheal ACC regardless of the treatment [1, 3, 7, 9]. In another review of 30 patients with ACC of the trachea and bronchi, the 5- and 10-year overall survival rates were higher at 84% and 70% with a median follow-up time of 59 months. Patients who underwent definitive surgical resection, regardless of the status of margins had much higher survival rates. Six percent of these patients had local recurrence, and 36% had distant metastasis. In one of the largest series, Gaissert et al reviewed 135 patients with tracheal ACC and showed a 52% and 29% 5 and 10-year survival, respectively, in resected ACC, but only a 33% and 10% 5 and 10-year survival in unresectable ACC of the trachea. In contrast to laryngeal ACC, tracheal ACC seems to be more commonly associated with poorer local regional control as demonstrated by Calzada et al with a 40% incidence of local recurrence. Negative surgical margins are difficult to obtain because of the relative inability to resect more than 6 cm of the trachea and the poor results associated with tracheal grafts. In our study, the patient had negative surgical margins [7-9]. In previous studies, it has been observed that patients with negative airway margins had statistically significant Disease free survival (DFS) rates.

5. Conclusion

ACC of trachea is an uncommon entity and can be easily confused with other functional causes of airway obstruction because of a similar clinical presentation. Complete surgical resection provides the patient with the best chance of prolonged survival or even complete remission. Therefore, it is important to be aware of the possibility of this entity presenting as a tracheal lesion with an emphasis on timely and accurate biopsy diagnosis leading to the situation of an early and appropriate treatment.

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