



Case Report

# Adenoid cystic carcinoma in trachea: A rare presentation

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## ABSTRACT

Primary tracheal tumors are rare. Adenoid cystic carcinoma of trachea is even rarer. It occurs as a polypoidal mass in trachea presenting with dyspnea and respiratory distress due to tracheobronchial obstruction. We report a case of young male presenting with dyspnea for some time. CT scan revealed mass in trachea obliterating its lumen. Histopathological examination confirmed the diagnosis.

## INTRODUCTION

Adenoid cystic carcinoma is a rare tumor of salivary gland origin, found mostly in head and neck region but has also been reported in breast, lacrimal gland of eye, lung, Bartholin's gland, trachea and paranasal sinuses.<sup>1,2</sup> Primary tracheal tumors are rare and constitute only two 2% of all respiratory tract tumors.<sup>1</sup> Adenoid cystic carcinoma is the second most common malignant tracheal tumor after squamous cell carcinoma.<sup>1,2</sup> These tumors should be considered because they often present as dyspnea or respiratory distress and

hence misdiagnosed and treated as asthma for months to years or sometimes until even with metastasis.<sup>3</sup> We report a case of young male presenting with polypoidal tracheal mass with complaints of dyspnea and respiratory distress as initial clinical presentation. Histopathological examination revealed it as Adenoid cystic carcinoma, a malignant tumor of salivary gland origin.

## CASE REPORT

A 31 year-old male presented with respiratory distress for 3 months. All the base line blood investigations were within normal limits. Bronchoscopy showed an intramucosal 2x2cm mass at mid trachea approximately 5 cm distal to vocal cord and 6 cm proximal to carina. CT scan revealed well

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defined heterogeneously enhancing broad based polypoidal soft tissue mass of 20x19x14.5 mm from posterior wall of trachea obliterating its lumen. (fig.1) Bilateral lungs have normal aeration. Operative findings revealed polypoidal mass measuring 2x2 cm attached to trachea over mid trachea with intact adventitia. Mass was obliterating around 80% of tracheal lumen. (fig.2) No palpable lymph nodes present. Grossly, tracheal tissue with an intraluminal mass attached to tracheal ring. Microscopically lesion revealed basaloid cells arranged in infiltrating cords, cribriform pattern along with some tubules. (fig.3) Tumor cells show moderate degree of atypia. Some pseudocyst with eosinophilic secretions noted.

## DISCUSSION

Adenoid cystic carcinoma of trachea was first described clinically and pathologically by Billroth in 1859. These are rare malignant tracheal tumors occurring in only 0.2 per 100000 people per year accounting for under 0.1% of the cancer deaths per year. These are second common tumor in trachea after squamous cell carcinoma.<sup>1-3</sup> Primarily these arise from the minor salivary glands and sero-mucinous glands of upper respiratory tract. Tracheal tumors are present mostly in lower and upper third with a tendency to originate at lateral and posterolateral wall near the junction of cartilaginous and membranous portions.<sup>1,2</sup> Squamous cell carcinoma is the most common pathology in smokers while adenoid cystic carcinoma is more prevalent among non-smokers.<sup>4</sup> Benign tumors are xanthogranulomas and pleomorphic adenoma.<sup>3,4</sup>

These tumors show equal sex distribution and tend to occur in the fourth and fifth decades of life.<sup>5</sup> Clinically they present with respiratory symptoms such as coughing, wheezing and dyspnea, hemoptysis, recurrent pneumonitis and are often treated for asthma for many time before they are correctly diagnosed.<sup>1,3</sup> In our case they presented as progressive dyspnea followed by respiratory distress for 3 months.

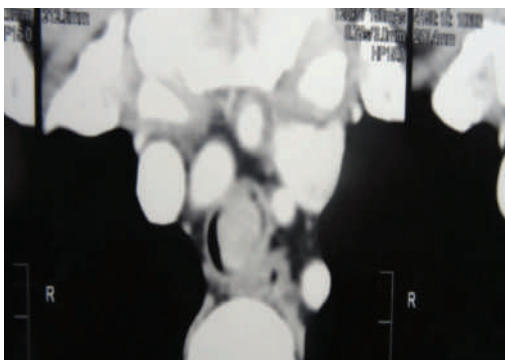
The radiological appearance of tumors can be classified as: intra-luminal, wall thickening, exophytic form. Endoscopic

evaluation reveals that the majority of lesions are bulky and obstructive. The main advantage of imaging is demonstration of tracheal wall thickening and extraluminal changes.<sup>4</sup> CT scan is useful and highly accurate in assessment of tumor location, invasion and metastasis.<sup>1</sup>

Pathologically they present as smooth polypoidal mass obliterating the airway lumen of trachea. They occasionally grow circumferentially or in annular pattern. They can spread along both submucosal and perineural planes which are responsible for local late recurrences.<sup>1,2</sup> They mostly have indolent behavior with a prolonged slow and insidious progression, in several years, despite its chances of local recurrences and distant metastasis.<sup>2</sup> However when it is associated with distant metastasis, survival is frequently less than 2 years.<sup>3,6</sup> Metastasis to brain, liver, kidney, skin, abdomen and heart have been reported.<sup>7</sup>

Adenoid cystic Carcinoma often invaded the cartilaginous plate of trachea, extending to lung parenchyma and even to hilar and mediastinal soft tissue. They show heterogenous growth patterns, where the neoplastic cells are arranged in cribriform pattern, tubules and solid nests. These tumour cells are small with scant cytoplasm and dark hyperchromatic oval to angulated nuclei. Mitotic figures are not frequent. Formation of pseudocyst filled with eosinophilic Periodic Acid Schiff positive material or basophilic granular material are noted. Perineural invasion is often noted. The cells of origin include intercalated ducts, myoepithelial cells, secretory cells, and pluripotent reserve cells. Immunohistochemistry markers and staining pattern vary depending upon tumour histogenesis. They express cytokeratin, vimentin, smooth muscle actin, p63, and GFAP. The surrounding matrix recapitulates a basement membrane like material staining with type IV collagen, laminin, heparin sulphate.<sup>5,8,9</sup>

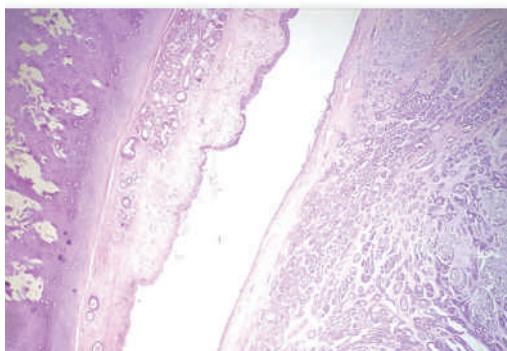
Recommended treatment consists of surgical resection with postoperative radiotherapy. However they have significant local invasion at the time of detection, making surgical resections more extensive.<sup>3</sup> Our patient is doing well and is



**Figure 1:** CT scan showing mass in tracheal lumen.



**Figure 2:** Photograph of trachea with polypoidal mass obliterating the lumen.



**Figure 3:** Low power view of tumour along with tracheal cartilage, mucous glands and respiratory epithelium. (HE stain X100).

on regular follow-up.<sup>10</sup>

## CONCLUSION

Tracheal adenoid cystic carcinoma presenting with respiratory distress may be misleading and hence correct diagnosis may be delayed. The publication of this article is to make aware about the unusual presentation of adenoid cystic carcinoma in trachea, which is slow growing, has multiple recurrences with late metastasis. Prompt diagnosis and treatment with regular follow up is needed.

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