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## Case Report

# Adenoid cystic carcinoma of trachea presenting as a medical emergency- A case report

Shrusty Advait Panchal Sonar<sup>1,\*</sup>, Anjali Mahajan<sup>1</sup>

<sup>1</sup>Dept. of Pathology, Lokmanya Tilak Municipal Medical College and General Hospital, Mumbai, Maharashtra, India



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### ABSTRACT

Adenoid cystic carcinoma is a chief histologic type of malignancy of minor salivary glands. Primary malignant Tracheal neoplasms are rare <0.1%, with Adenoid Cystic Carcinoma being very rare. It arises from mucous secreting cells of salivary glands and upper respiratory tract. After Squamous Cell Carcinoma it is second most common Primary tracheal malignancy. Diagnosis can be done by Computed Tomography or Magnetic Resonance Imaging. But histopathology remains gold standard of diagnosis.

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## 1. Introduction

Adenoid cystic carcinoma (ACC) of trachea is not very common with incidence of 0.1 to 0.26 per 1 lakh people.<sup>1-3</sup> Adenoid Cystic Carcinoma has an indolent course. More frequently it involves Minor salivary glands than major. Other sites include tongue, Paranasal sinuses, palate, nasopharynx, lacrimal glands, and external auditory canal. Also may originate in the tracheobronchial tree, oesophagus, lungs, breast, prostate, Bartholin's glands, cervix and vulva.<sup>4</sup> Diagnosis is usually delayed as the symptoms may mimic other conditions.<sup>5</sup> It may lead to luminal narrowing causing airway obstruction, which may be life threatening requiring emergency intervention.

## 2. Case History

A 65 year old male presented to the emergency with breathlessness. Emergency Tracheostomy was performed. Tracheal mucosal thickening was noticed during the procedure in view of which tracheal biopsy was done.

Patient's relatives gave history of him suffering from Dyspnoea and of Noisy breathing since 4 months.

History of tobacco chewing since 10 years.

Post procedure CT Neck revealed Concentric mucosal thickening in Post cricoid region of trachea which was causing luminal narrowing.

### 2.1. On Histopathological examination

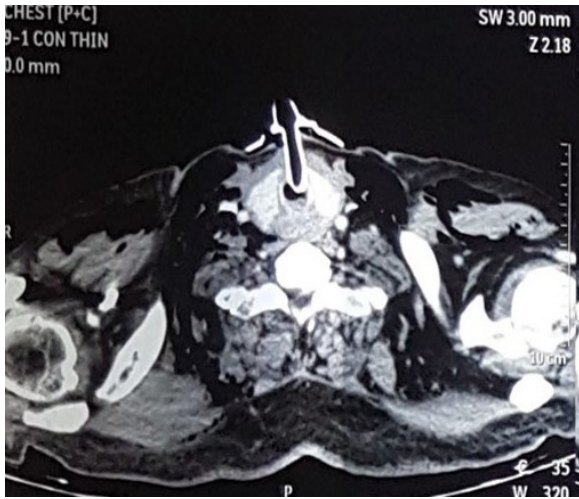
Two grey white bits of tissue measuring 1×0.5 and 1.5×0.8cm were received. Tissues were processed. Three to five microns sections were taken and stained with Haematoxylin and Eosin.

On microscopy, of H&E sections showed typical features of Adenoid cystic carcinoma. Moderately Uniform cells with hyperchromatic angular nuclei with scant eosinophilic cytoplasm. The cells were arranged in tubule, nest in cribriform pattern. The spaces were filled with homogenous eosinophilic material.

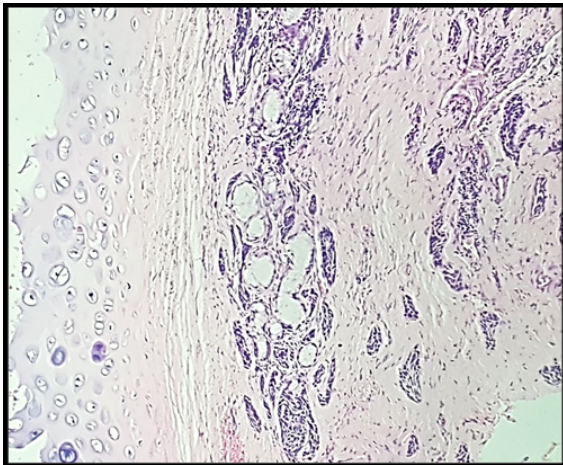
Nuclear atypia was moderate. No necrosis was seen. Mitotic figures were not seen.

\* Corresponding author.

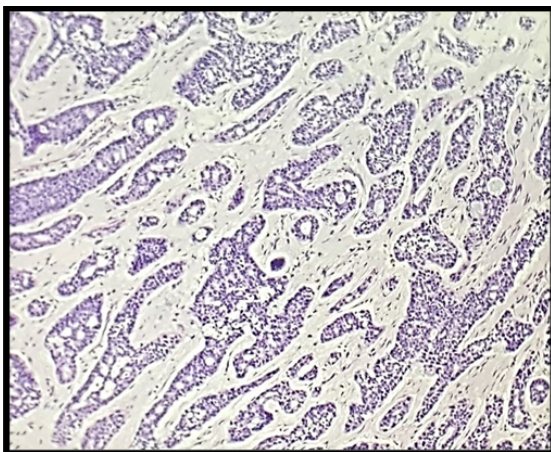
E-mail address: [shrustypanchal92@gmail.com](mailto:shrustypanchal92@gmail.com) (S. A. P. Sonar).



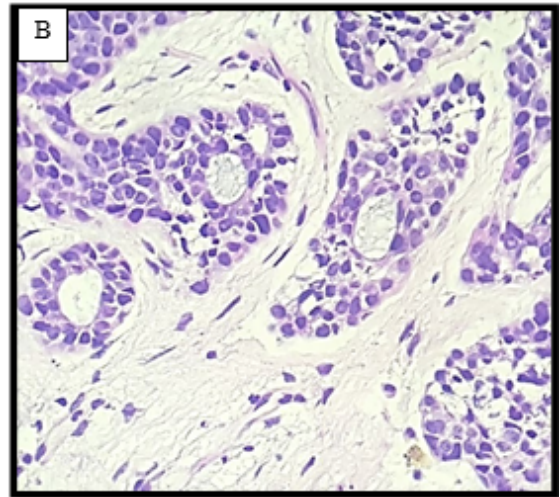
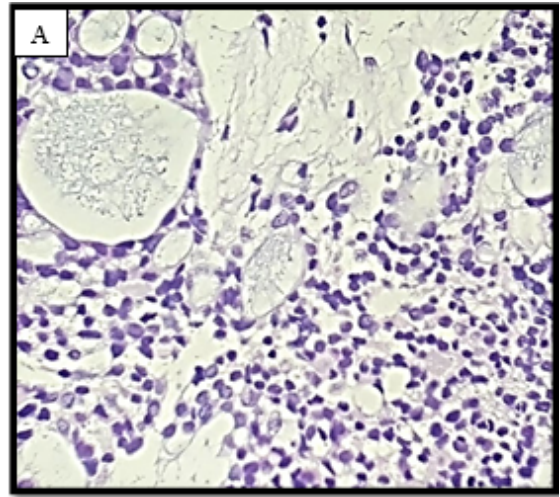
**Fig. 1:** CT findings: Concentric mucosal thickening in post cricoid region.



**Fig. 2:** 100x-shows tracheal cartilage and tumour cells arranged in nest, tubules and in cribriform pattern.



**Fig. 3:** 100x-shows tumour cells arranged in nests and tubules. Good amount of extracellular matrix can be seen within cyst like spaces formed in several nests.



**Fig. 4: a and b:** 400x -Moderately Uniform cells with hyperchromatic nuclei and little cytoplasm. The tumor cells have small to inconspicuous nucleoli. Finely dispersed chromatin seen.

### 3. Discussion

This case demonstrates airway compromise as a late presentation of tracheal adenoid cystic carcinoma. Adenoid Cystic Carcinoma is relatively slow growing with an increased infiltrative nature. The growth can be nodular or smooth lateral spread causing concentric narrowing of airway. Distal one third of trachea is the most frequently affected site. Proximal trachea and the larynx are less involved.

ACC can occur at any age but is more commonly seen in the fourth and fifth decades of life. Males and females are almost equally affected.<sup>6</sup> Symptoms are usually nonspecific including cough, dyspnoea and at times haemoptysis. These symptoms overlap with other much common conditions like asthma leading to misdiagnosis. Tumor spread occurs by direct extension submucosally or via perineural invasion. Distant metastasis can also occur most frequently to lungs

and occasionally to liver, brain and bony skeleton. Local recurrence is also seen.

They are usually solitary and nodular not more than 4 cm in size and have a firm yellow-white cut surface.<sup>6</sup>

The term “cylindroma” was coined originally for ACC due to its microscopic appearance of cylinders of cells with hyaline stroma.

The tumor is composed of a biphasic cell population consisting of ductal and basal or myoepithelial differentiation.<sup>6</sup>

Cells have angulated hyperchromatic nuclei and clear or slightly eosinophilic cytoplasm which is scant in amount.

The major histological subtypes include cribriform, tubular and solid forms. The cribriform pattern is commonly seen. Basaloid cells forming islands which are surrounded by different sized cyst-like spaces. These cyst like spaces are not true glandular lumina. The tubular pattern has cells placed in nests amidst eosinophilic stroma of variable amounts. The solid variant shows aggregates of basaloid cells with no tubule or pseudo-cystic forms.<sup>4</sup> The solid variant has the worst prognosis as it is high grade, much aggressive and shows distant metastasis.

Immunohistochemical stains are usually not required for the diagnosis but can be helpful in certain cases especially to diagnose solid or tubular ACC's. Immunostaining of basal cells is generally positive for myoepithelial markers like p63, S-100, Calponin. Also shows positivity with vimentin. Whereas ductal cells display positivity for CK7, EMA, CK5/6 and ckit.<sup>7</sup>

Surgery is the main mode of treatment along with radiotherapy.<sup>2</sup> The 5 year survival and 10 year survival rate of patients with tracheal ACC ranges from 52 to 79% and 27 to 51% respectively; the survival being longer in patients with complete resection, negative margins and also in patients who receive adjuvant radiotherapy.<sup>8</sup>

#### 4. Conclusion

Distal Tracheal masses should be remembered as rare but serious cause of progressively worsening difficulty in breathing.<sup>1</sup> ACC is an infrequent tumor and due to its low occurrence it may be missed. Early diagnosis and treatment is very essential for better prognosis.

#### 5. Abbreviations

ACC- Adenoid cystic carcinoma,

#### 6. Conflict of Interest

The authors declare no relevant conflicts of interest.


#### 7. Source of Funding

None.

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#### Author biography

**Shruty Advait Panchal Sonar**, Resident  <https://orcid.org/0000-0002-0275-6085>

**Anjali Mahajan**, Associate Professor

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