

# AN UNUSUAL CAUSE OF STRIDOR "ADENOID CYSTIC CARCINOMA OF TRACHEA"

Muhammad Mudassir Shafiq\*, Kausar Rehana\*,

\*Department of Pulmonology,  
Pakistan Institute of Medical  
Sciences, Islamabad-Pakistan

Address for correspondence:

**Kausar Rehana**

Department of Pulmonology,  
PIMS, Islamabad-Pakistan

Email: seasaw701@gmail.com

## ABSTRACT

Previously called cylindroma or adenocystic carcinoma, Adenoid cystic carcinoma (ACC) of the trachea is rare, it represents less than 1% of all respiratory tract cancers. It is a slow-growing cancer, with prolonged clinical course. Most patients present with dyspnea and wheeze/stridor, often mimic those of asthma. Surgical resection is the mainstay of treatment combined to adjuvant radiotherapy because of close resection margins. We report a case of primary ACC of trachea: a 45 years old female presented with dyspnea and stridor having this tracheal tumor treated by surgical resection and reconstruction with end to end anastomosis followed by adjuvant radiotherapy. At 6 months follow-up, our patient shows no evidence of disease with negative histological findings.

**Keywords:** Adenoid cystic carcinoma; surgical tracheal resection; adjuvant radiotherapy

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## SUMMARY

**A** 45 year old house wife presented to Pulmonology OPD with complaint of dyspnea from last 1 year. She was in normal state of health about 1 year back when she developed cough with high grade fever and presented to local hospital. There she was labeled as a case of "Community Acquired Pneumonia" and got discharged on oral treatment after stay of 3 days. She felt some improvement but after 2 weeks of that event she again developed severe dyspnea that was associated with fever, wheezing and orthopnea. With these complaints she landed in a private hospital and labeled as a case of "Bronchial Asthma" by a pulmonologist and was put her on treatment including inhalational drugs.

After 6 months, beside that she was on treatment and well compliant, she became more and more dyspnoeic and got admitted with diagnosis of "Acute severe asthma" in a tertiary care hospital. There, she was managed in medical ICU, and diagnosis was reviewed as some "upper respiratory tree issue vs obstruction". Contrast-enhanced computed tomography (CECT) chest was ordered there and proved to be having some mass lesion that compressing trachea (Figure 1). With this report she was referred here in our OPD. She got admitted here as she was markedly tachypnic and in distress. She had no premorbid,

never smoked or drunk and had no pet exposure. She belongs to a middle class family and had no h/o nasal symptoms, GI, GU, CNS related issues.

She was tachypnic and was in respiratory distress using accessory muscles, had obvious stridor. Chest examination showed central trachea, stridor all over the chest with areas of bronchial breathing and increase vocal resonance in lower chest B/L. After stabilizing patient her bronchoscopy was planned that showed "Intratracheal growth that obstructing almost all of its lumen" (Figure 2). Multiple Biopsies were taken and sent for H/P. She was managed with antibiotic steroids and high flow oxygen. While she was on symptomatic treatment her biopsy report was received. Reported as "Adenoid cystic carcinoma of trachea" (Figure 3).

Thoracic surgeon was consulted and he demands three dimensional "CT scan neck and chest with reconstruction images". So, his demands were fulfilled and these were CT images. Incidental findings in this CT was "Areas of pneumonitis B/L lower zones likely to be consequence of retained secretions" (Figure 4). So plan of surgical resection was postponed till her infection was resolved and she was put on antibiotics. She improved clinically, starting to maintain saturation at room air and CXR was also showed marked resolution of pneumonitis. After 1

week of antibiotics she was again sent for thoracic surgeon review and finally admitted there. Her tracheal mass resection with reconstruction was done successfully and was discharged.

She remained well and follow up her pulmonology OPD and in thoracic surgery OPD. She was

advised to repeat her CT scan neck and chest after 1 month that shows significant recanalization of diseased segment (Figure 5). After this CT, radiotherapy treatment was decided for her and for this referred her to Oncology unit, where treatment for here was started and she successfully completed her treatment.

Figure 1:



Figure 2:



Figure 3:

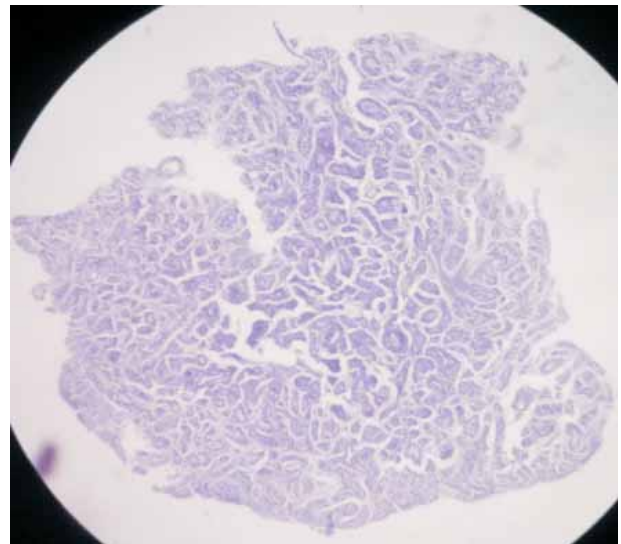


Figure 4:

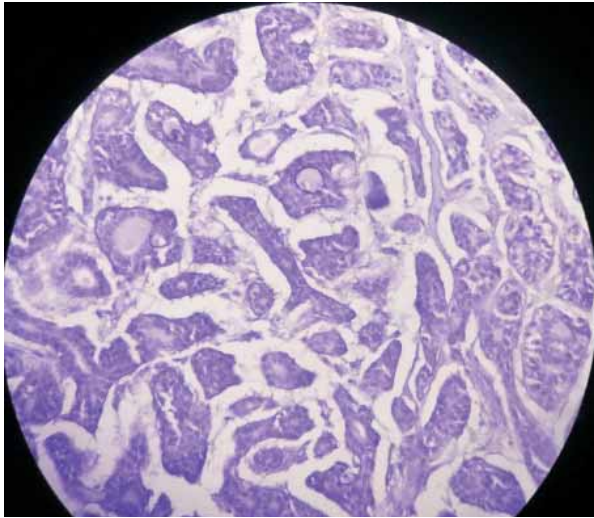


Figure 5:



Figure 6:

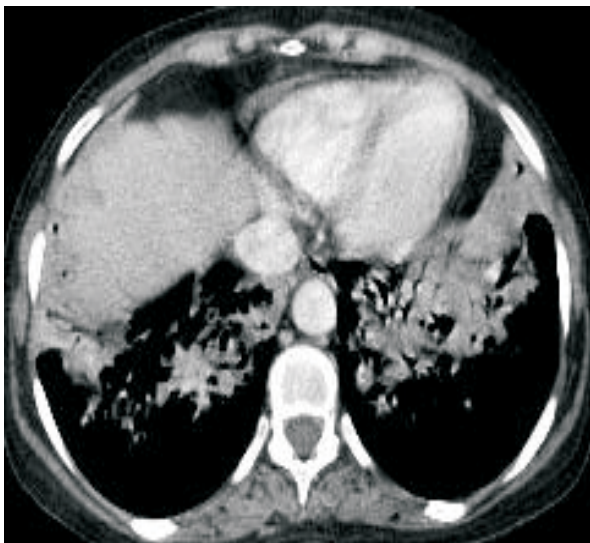


Figure 7:



## DISCUSSION

ACC of the trachea is a relatively rare occurrence. The overall incidence of tracheal tumors is under 0,2 per 100,000 people per year ;<sup>1</sup> ACC is the only 10%.<sup>2</sup> ACCs have been reported without gender predilection in patients usually in the fifth decade of life; our patient is 45year female and the incidence of ACC is not affected with smoking.<sup>3</sup> Patients with ACC usually present with symptoms such as coughing, wheezing and dyspnea and are often treated for asthma, as in our case, for months to years before being correctly diagnosed .<sup>1</sup> ACC is a nonencapsulated tumor and spreads

most commonly by direct extension, perineural invasion or submucosal, in longitudinal and transverse planes.<sup>4</sup> More than 50% of patients with ACC of trachea have hematogenous metastases. Pulmonary metastases are the most common and usually remain symptoms free for many years.<sup>3</sup> CT scan and MRI is used to define extent and infiltration if any.<sup>3,5</sup> We use CECT Scan neck with reconstruction images for this purpose.

Treatment options include surgical resection alone, radiotherapy alone; or in combination. The ideal treatment of ACC is primary resection and reconstruction with end-to-end anastomosis when possible (as we did). Surgical resection has

been thought to be the most favourable procedure to control localized lesions. The median survival time of surgically resected patients was reported to range from 7,5 years to 118 months.<sup>6</sup> Due to infiltrative nature, ACCs are often incompletely resected but in our case complete resection was achieved.

The role of adjuvant radiotherapy remains uncertain. In adjuvant situations the overall survival rate at 5 and 10 years were respectively in three major series 66-79% and 51-57% .<sup>7-9</sup> With limitations of randomized comparison, it stills seems reasonable to recommend adjuvant radiotherapy post resection, and certainly for those patients in whom the pathologic examination identifies positive margins. One month period is recommended after surgery and it may be useful to perform a bronchoscopy or CECT to ensure healing. Chemotherapy does not appear to have a role in the treatment of ACC of the trachea .<sup>1</sup>

## CONCLUSION

Adenoid cystic carcinoma is a rare primary tracheal malignancy. Patients usually present with stridor/wheeze. This is commonly misdiagnosed as asthma. Surgical resection with reconstruction followed by adjuvant radiotherapy is widely recommended protocol for treatment of this tumor and provides improved survival.

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