

Case Report



Adenoid Cystic Carcinoma of Trachea: Two Case Report

Islam U. Chowdhury¹, Md. Mofazzal Sharif²,
Md. Shahidul Islam³, Afzal Hossain⁴, Ashiul Amin⁵, Md. Abdul Hai⁶.

Abstract

Primary carcinoma of trachea also known as adenoid cystic carcinoma, adenocyst, malignant slyndroma. This tumor mostly occurs in the salivary gland (minor), lacrimal gland, trachea, paranasal sinuses and others. And also can occur in female reproductive tract, Bartholin gland and cervix. Carcinoma trachea is one of the very rare cases. Adenoid cystic carcinoma (ACC) of trachea is one of the very rare cases. This type of carcinoma is very slow growing cancer. Usually patients present with dyspnoea, asthma or bronchitis. After confirmed diagnosis 1st indication is surgery, if patient is not fit for surgery, can go to the laser ablation then Radio therapy. Our two cases of adenoid cystic carcinoma of trachea are, one is 30 years old female patient, her initial diagnosis was May, 2012 and another one 48 old male patient with history of smoking, his initial diagnosis was on July 2003. Both the patients are still alive. Now the female patient has bilateral pulmonary metastasis from September 2017. After treatment carcinoma of trachea can be metastasised to lung, brain, liver or others organs. Usually it happened after 12-15 years from initial diagnosis. However carcinoma of trachea has good prognosis if early diagnosed and treated.

Keywords: Trachea, Adenoid cystic carcinoma, Radiotherapy, Chemotherapy.

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Introduction

The incidence of tracheal carcinoma is less than 0.2-0.26/100000 person per year. And 0.1-0.4% of all malignant disease.¹⁻³ Adenoid cystic carcinoma is not associated with smoking. Being unencapsulated carcinoma it mostly spread by direct metastasis. Only 20% of adenoid cystic carcinoma spread to lymph nodes. This incident may be related to part of the hard palate and paranasal sinuses, because these areas frequently produce lymph node metastasis.^{4,5} The tumours may be histologically classified into tubular, cribriform, and solid subtypes. The tumours are usually low grade but may occasionally undergo high-grade transformation.⁶ On chest CT, these tumors have a notable tendency toward submucosal extension, typically manifest as intraluminal mass of soft-

tissue attenuation with extension through the tracheal wall or diffuse or circumferential wall thickening of the trachea, a soft-tissue mass filling the airway and homogeneous mass encircling the trachea with wall thickening in the transverse and longitudinal planes. The longitudinal extent of the tumors is greater than their axial extent and the tumors usually involve more than 180° of the airway circumference.^{4,6,7} Currently, there are no known methods to prevent Tracheal Adenoid Cystic Carcinoma occurrence. Regular medical screening at periodic intervals with blood tests, scans, and physical examinations, are mandatory, due to its metastasizing potential and possibility of recurrence. Often several years of active vigilance is necessary.⁸⁻¹⁰

1. Consultant Medical oncologist, Ahsania mission cancer and General Hospital, Dhaka, Bangladesh.
2. Associate Professor, Department of Radiology & Imaging, KhwajaYounus Ali medical college and Hospital, Enayetpur, Sirajganj, Bangladesh.
3. Associate Professor, Department of Oncology, KhwajaYounus Ali medical college and Hospital, Enayetpur, Sirajganj, Bangladesh.
4. Resident, Department of Oncology, KhwajaYounus Ali medical college and Hospital, Enayetpur, Sirajganj, Bangladesh.
5. Medical officer, Cancer unit, Gonoshasthya Somaj Vittik Medical College & Hospital, Savar, Dhaka, Bangladesh.
6. Professor & Director of Bangladesh Cancer Society, Bangladesh.

Correspondence: Dr. Islam U. Chowdhury, Consultant Medical oncologist, Ahsania mission cancer and General Hospital, Dhaka, Bangladesh, Email: chowdhuryis@yahoo.com

Case Reports

Case 1

A 30 year old female patient, non-hypertensive, non-diabetic, occasionally complained anorexia and respiratory distress. On April, 2012 MRI of neck report showed irregular soft tissue mass in the trachea and right paratracheal region occluding almost lumen of the trachea. Histopathology report (tissue from tracheal growth) revealed adenoid cystic carcinoma on May, 2012. After diagnosis, surgery was done and post operative finding was a fleshy mass arising from posterior wall of trachea and extends from 2nd to 5th tracheal ring. Then she received Radiation therapy (anterior and posterior chest wall) with 44 Gy in 22 fractions from May, 2012 to June, 2012. Then she was on regular follow up and was well till December 2016 with no evidence of disease revealed at bronchoscopy (Figure 1). On September 2017, she complained respiratory distress, diffuse chest pain, which was aggregated with inspiration, nausea, vomiting and sleep disturbance and right hypochondriac pain. No organomegaly, palpable lymph node was detected. On September 2017 contrast enhanced CT and PET-CT showed bilateral pulmonary metastases, focal enhancing tissue in right side of trachea invading upper part of the oesophagus, extending from C5-C6 level to upper part of D1 with focal enhancing soft tissue in posterior wall of nasopharynx invading both medial pterygoids, at vallecula and level II lymph nodes (Figure 2 and 3). CT guided FNAC from lung showed metastatic lung carcinoma one on September 2017. Then we started chemotherapy with Docetaxel 80 mg and carboplatin 450 mg, in 3 weeks interval. She received 6 cycle chemotherapy with GCSF support. Now patient is symptomatically better than before but not completely free from lung metastasis till 2019.

Case 2

Our second patient is 67 years old male with diabetes, history of smoking, diagnosed as a case of adenoid cystic tumour of trachea on 2003 when he was Middle aged. After laser ablation and post Radiation therapy, he was suffering frequently from cough and breathlessness. As condition of patient did not improve and condition gradually deteriorated he was advised for bronchoscopy (Figure 4). Biopsy report showed adenoid cystic carcinoma of trachea (at the level of 10th -11th tracheal ring). He was again advised for laser ablation of the tumour. As he was not fit for surgery, due to involvement of the whole thickness of the trachea, he received only one course of chemotherapy with Methotrexate and cisplatin. After treatment patient was improved for about 1 year. Then in mid of 2004 again suffered from fever, cough and haemoptysis. Again bronchoscopy was done, result showed recurrence of the disease (Figure 5) and he received laser ablation. After 5 months follow up bronchoscopy was done and found recurrence again (Figure 6), then 3rd time laser ablation was done. In next follow up, 4th time recurrence was found and he received Radiotherapy with 40 Gr in 20 fractions. After completion of RT patient's condition was not improved and residual remained. Then 2nd time he received Radiotherapy

with 61 Gr in 33 fractions, (May 2008 to June 2008). Then in 2 years of regular follow up the patient was well. Again 5th time recurrence occurred and he got 3rd time radiotherapy by 45 Gr radiation in 15 fractions (May 2010 to June 2010). Now he is in regular follow up, disease is in control and he is well till now (2019) (Figure 7).

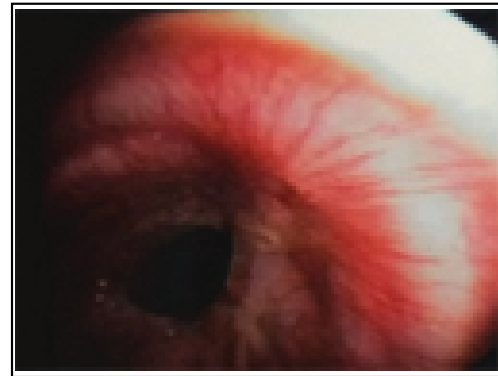


Figure 1: Endobronchial endoscopy showing no residual disease

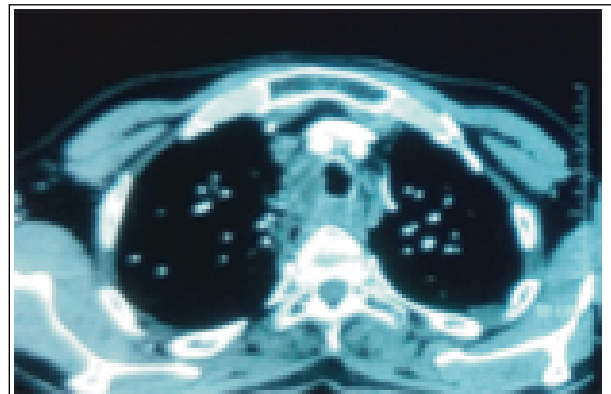


Figure 2: Contrast enhanced CT scan of chest showing enhancing soft tissue around right lateral aspect of trachea.

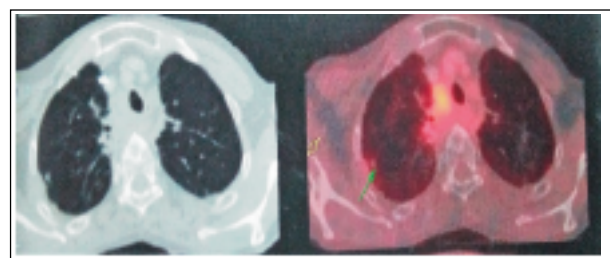


Figure 3: PET-CT image showing FDG avid area in right lateral aspect, posterior wall of trachea and pulmonary metastases.

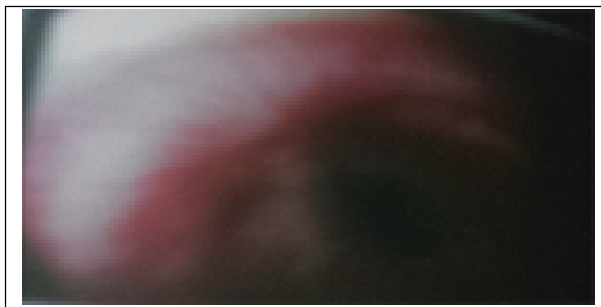


Figure 4: Endobronchial endoscopy showing recurrence of disease along anterior wall.

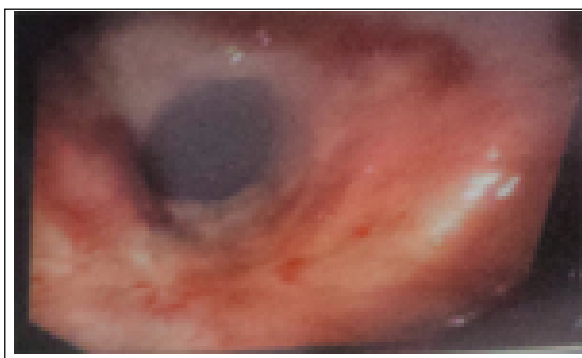


Figure 5: Repeat endobronchial endoscopy in mid of 2004 showing recurrence of disease along anterior wall.

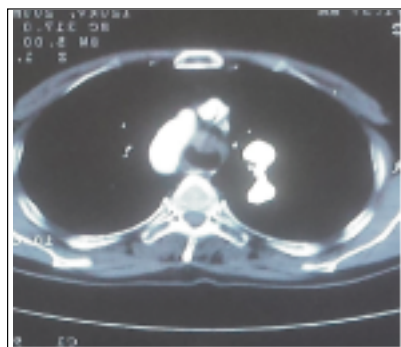


Figure 6: Contrast enhanced CT scan of chest showing enhancing soft tissue in anterior aspect of trachea.

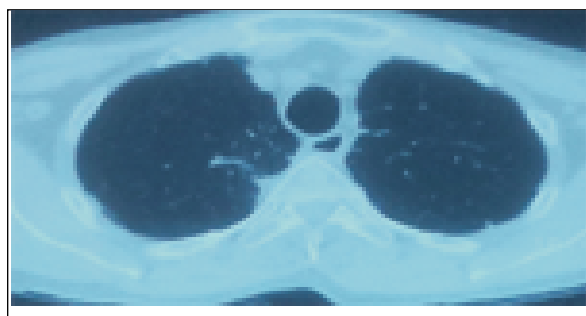


Figure 7: CT scan of chest in lung format showing no residual disease (in 2019).

Discussion

Previously called cylindroma or adenocystic carcinoma, ACC of the trachea is a relatively rare occurrence. The incidence of tracheal tumours is less than 0.2 per 100,000 people per year, only 10% of these cases are ACC.^{1,8} ACC's have been reported without sex predilection in patients in the fifth decade of life; our patients are 49 and 50 years old; and smoking does not affect the incidence of ACC.^{3,4} Patients with ACC usually present with symptoms such as coughing, wheezing and dyspnoea and are often treated for asthma for months to years before being correctly diagnosed, as in our second patient. ACC is none capsulated tumour, it spreads most commonly by direct extension, submucosal or perineural invasion, in transverse and longitudinal planes.^{4,5} Lymphatic spread is uncommon. More than 50 % of patients with tracheal ACC have haematogenous metastases.^{3,6} Pulmonary metastases are the most common and can remain asymptomatic for many years.^{5,6} Metastases to the brain, bone, liver, kidney, skin, abdomen; and heart have also been reported.³ On CT, the tumour infiltration in the submucosa manifests as an intraluminal mass of soft-tissue attenuation with extension through the tracheal wall, and a diffuse or circumferential wall thickening of the trachea, a soft-tissue mass filling the airway, or a homogeneous mass encircling the trachea with wall thickening in the transverse and longitudinal planes. MRI can better define the extent of the submucosal infiltration and local mediastinal invasion than CT that may influence resectability.¹⁻³ Treatment options include surgery alone, radiation therapy alone; or a combination. The ideal treatment of ACC is primary resection and end-to-end anastomosis when possible. A surgical resection has been thought to be the most favourable procedure to control localized lesions. When the tumour is considered unresectable due to its local extension, exclusive radiotherapy is given. Conformal radiation therapy is currently recommended. The role of post-operative adjuvant radiotherapy remains uncertain.^{7,8}

Conclusion

ACC is a very rare primary tracheal malignancy. This disease is commonly misdiagnosed as COPD and asthma. Surgical resection followed by radiotherapy is widely recommended protocol for treatment of localized tracheal tumours. Patients with unresectable disease, can be benefited with Laser ablation, chemotherapy and Radiotherapy.

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