

Primary Adenoid Cystic Carcinoma of Trachea: Managed by Tracheal Resection and Reconstruction

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ABSTRACT

Tracheal tumors are infrequent and account for less than 1% of all malignancies of this area¹. We are reporting a case of a 43 years old female who presented with shortness of breath, tiredness, dry cough and hemoptysis for one year. CT scan demonstrated a soft tissue density mass in the trachea causing narrowing of the tracheal lumen. Tracheal resection and primary tracheo-tracheal end to end anastomosis was performed. Per operatively tumor was found in the mid of trachea, 4.5cm in length craniocaudally. Biopsy report showed features consistent with Adenoid Cystic Carcinoma. The patient is in regular follow up for last 14 months and remains well.

KEY WORDS: Tracheal Malignancy, Adenoid Cystic Carcinoma, Tracheo-Tracheal Anastomosis.

This article may be cited as: Soomro NH, Khan I, Ansari M, Zafar AA. Primary Adenoid Cystic Carcinoma of Trachea: Managed by Tracheal Resection and Reconstruction. J Liaquat Uni Med Health Sci. 2016;15(04):213-6. doi: 10.22442/jlumhs.161540495

INTRODUCTION

Primary tumors of the trachea are rare and are the cause of <0.1% of the cancer deaths per year¹. Smoking is the most commonly associated risk factor². Squamous cell carcinoma is the most common type followed by Adenoid Cystic Carcinoma and both of them constitute roughly 75% of all tumors of the trachea³. Salivary gland is the most common site of occurrence of Adenoid cystic carcinoma whereas trachea, lacrimal glands and eyes are other possible locations. It occurs most frequently in the salivary glands and not related to smoking². They occur equally in men and women between the ages of 40 and 60 years⁴. It spreads along the lining of the trachea, grows slowly and is associated with prolonged survival. Complete surgical resection can offer a better prognosis and increases chances of survival. Surgery, in combination with radiotherapy or chemotherapy is the usual mode of treatment. In patients who need palliative therapy only, laser removal of the intra tracheal tumor can be attempted. We are presenting a case of primary tracheal adenoid cystic carcinoma; treated by surgical resection of the lesion and end to end tracheo-tracheal anastomosis.

CASE REPORT

A 43 year old female presented with shortness of breath, tiredness, dry cough and hemoptysis for one year. These symptoms were present even at rest. She was on inhalers but there was no improvement in dyspnea. She denied any history of smoking. On examination, her vitals were within normal limits. Chest

exam was unremarkable except for audible stridor. The rest of the systemic exam as well as all hematologic laboratory investigations were normal. The Chest X ray did not show any pulmonary lesion. CT scan showed intraluminal soft tissue density mass lesion in the trachea at the level of sternoclavicular joint measuring 2.6 x 2.2 x 3.6 cm, 1.5 cm above the carina, almost obstructing lumen of the trachea. Anteriorly, it was abutting the right subclavian artery with intervening fat planes and posteriorly it was abutting the esophagus.

After preoperative work up, she was planned for tracheal resection and anastomosis. She was anaesthetized by using single lumen ETT. It was placed above the tumor and low tidal volume hand ventilation performed till the tumor was resected out completely. Per operatively tumor was found in the mid of trachea about 2 cms above the carina and 5.5 cm of the trachea was resected along with the tumor. Distal part of the trachea was resected first and a new single lumen ETT was placed in left main bronchus to ventilate left lung and then the proximal part of trachea was resected. After that, distal tracheal, carinal and hilar mobilization was performed including inferior pulmonary ligament. Primary tracheo-tracheal end to end anastomosis was done. Tumor specimen was sent for histopathology. Biopsy report showed features consistent with Adenoid Cystic Carcinoma and margins were tumor free. The case was discussed at the tumor board meeting and the panel of oncologists did not recommend adjuvant radio or chemotherapy. The patient is on regular follow up for last 14 months and has been doing well.

FIGURE I: PEROPERATIVE PICTURE SHOWS TRACHEAL TUMOR, RESECTED PROXIMALLY AND DISTALLY (BLACK ARROW), THE ETT TUBE PASSED IN LEFT MAIN BRONCHUS (YELLOW ARROW) TO VENTILATE LEFT LUNG

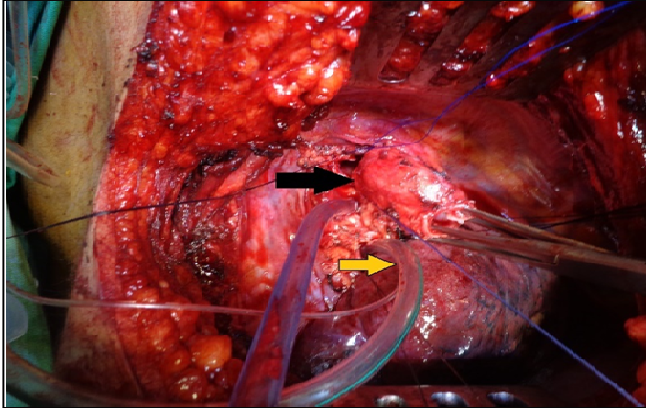


FIGURE II: RESECTED TRACHEA WITH TUMOR, APPROX. 5.5 CM AFTER CUTTING THE MEMBRANOUS WALL OF TRACHEA



FIGURE III: PREOPERATIVE CT SCAN CHEST WITH CONTRAST SHOWS TUMOR IN THE TRACHEA, ALMOST COMPLETELY OBLITERATING THE TRACHEAL LUMEN

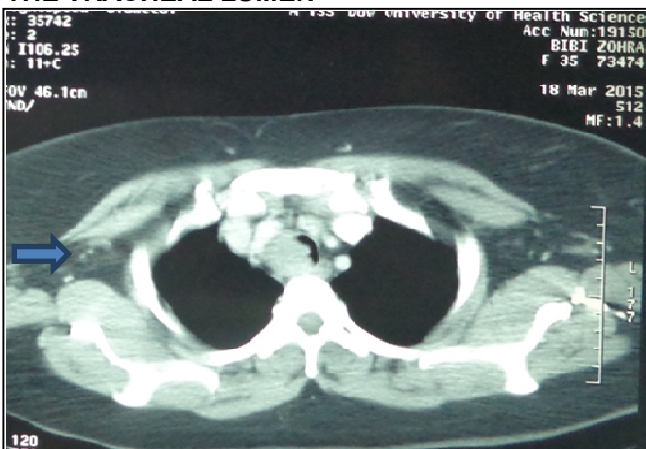
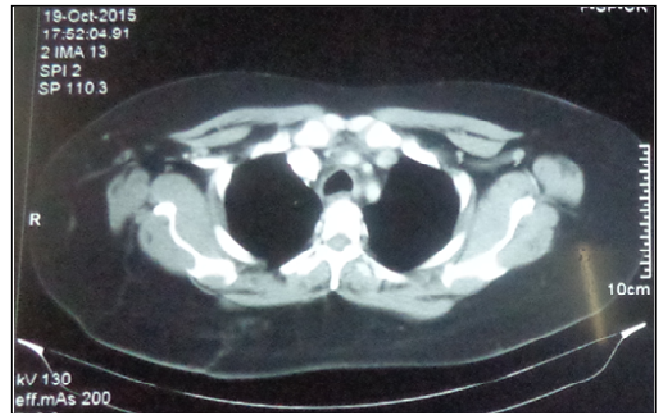


FIGURE IV: POST-OPERATIVE FOLLOW UP CT SCAN AFTER 3 MONTHS SHOWING RECONSTRUCTED TRACHEA WITH PATENT LUMEN



DISCUSSION

According to National Cancer Institute (NCI) surveillance approximately 2.6 per million new cases of primary carcinomas of trachea are reported every year⁵. These tumors make up only 2% of all upper respiratory tract tumors⁶. Tracheal tumors are usually malignant in adults and benign in children. ACC of the trachea is a rare neoplasm originating from submucosal glands of the tracheobronchial tree being predominantly a malignant tumor of the salivary glands and most common in the parotid gland representing about 10% of head and neck tumors⁷. Malignant tumors of the trachea occur more frequently than benign tumors and the most common benign tumors of trachea includes chondroma, hemangioma and papilloma. Among the common primary malignant tumors of trachea, squamous cell carcinoma is the most common, followed by adenoid cystic carcinoma and carcinoid tumor.

Dyspnea and shortness of breath on exertion, cough, hemoptysis and change in voice quality are common presenting symptoms¹. Wheezing becomes a prominent symptom as airway narrowing occurs and presence of stridor indicates significant compromise of the airway. These patients are often mistaken as having asthma and continue to be treated with bronchodilators and steroids for prolonged periods⁸ same as our patient treated initially. The mean duration of symptoms with adenoid cystic carcinoma was 12 months as observed by Regnard et al⁹.

The diagnosis is delayed until the tumor is identified via CT or bronchoscopy, because tracheal tumors are located in relatively blind fields in chest radiography. Chest radiogram reveals abnormalities in only 1/3 of the patients and laryngoscopy in only 25%, therefore airway CT or bronchoscopy should be performed, for patients, experiencing diagnostic problems¹⁰.

Tracheal squamous cell carcinoma (SCC) mostly occur in men approximately 90%¹, primary tracheal Adenoid Cystic carcinoma is found in men and women with almost equal frequency with peak incidence in fourth to sixth decade of life³, our patient belonged to same age group. It is not related to smoking. However it's a low malignancy tumor and because of its low potential for growth and delayed metastasis, it poses a diagnostic and therapeutic challenge.

According to Yang et al, resected tumor size and tracheal lengths tended to be smaller ($28.2 \pm 7.6\text{mm}$) as compared to our reported case¹. The most common site of tumor is in upper part of trachea, in contrast; we are reporting a tumor in mid part of trachea.

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 favorable procedure to excised localized lesions. The complete resection rates in reported cases ranged from 42 to 57%¹¹. Negative surgical margins are difficult to obtain because of the relative inability to resect more than 6 cm of the trachea, and thus are prone to recur locally¹². However, previous studies have found that overall survival rates of completely and incompletely resected ACCs are not significantly different, even when a residual tumor was left behind after surgical resection^{13,14}. Maziak DE reported five-year cumulative survival rates for patients with complete and incomplete resection as 82% and 77% respectively¹⁵. In contrast Gaissert HA et al showed statistically significant longer disease free survival with negative airway margins¹⁶. This lack of census among different studies regarding significance of tumor free margin is probably due to low numbers and inadequate follow-up. ACC seems to possess radio sensitivity, most tumors respond to radiotherapy which often results in long periods of remission for patients treated with radiotherapy alone¹⁷.

CONCLUSION

Patients with tracheal tumors may present with hemoptysis, dry cough, dyspnea, chest pain and weight loss, therefore any such patient not responding to bronchodilators should be evaluated for possible neoplastic lesion of trachea. Complete surgical resection offers the patient with the best chance of cure and increased survival rates.

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