Tracheal adenoid cystic carcinoma: A reported case and review of literature

Case Article

Omar Iskanderani

Department of Radiation Oncology, Faculty of Medicine, King Abdulaziz University, Jeddah, Saudi Arabia.

ABSTRACT

Introduction: Tracheal adenoid cystic carcinoma (ACC) is a rare but distinctive subtype of tracheal cancer. Surgery remains the primary treatment, but optimal postoperative radiotherapy regimens are still being explored to minimize local recurrence. This is a case of ACC of the trachea. An individualized radiotherapy regimen was meticulously planned after surgery. This tailored approach significantly impacted the patient's outcomes.

Case Report: A 49-year-old female with history of progressive dyspnea and cough that was unresponsive to conventional treatments over 6 months. Flexible bronchoscopy revealed an obstructing mass in the upper trachea, and biopsy confirmed the diagnosis of adenoid cystic carcinoma. Imaging ruled out distant metastases. The patient underwent tracheal sleeve resection, followed by adjuvant radiation therapy using Volumetric Modulated Arc Therapy (VMAT) and 6-megavoltage (MV) radiography. The patient tolerated the treatment well, with no significant acute or late toxicities. Follow-up imaging studies have shown no evidence of recurrence at 6 years post-treatment.

Conclusion: Tracheal ACC is a rare but distinct entity that requires prompt recognition and multimodality management for optimal outcomes.

Key Words: Adenoid cystic carcinoma, malignancy, trachea.

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Corresponding Author: Omar Iskanderani, MSc, Department of Radiation Oncology, Faculty of Medicine, King Abdulaziz University, Jeddah, Saudi Arabia, Egypt. **Tel.:** +966555556995, **E-mail:** oflskanderani@kau.edu.sa

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INTRODUCTION

Tracheal adenoid cystic carcinoma (ACC) is a rare but distinctive subtype of tracheal malignancy, accounting for 10% of all tracheal malignancies[1,2]. Despite its rarity, ACC of the trachea poses unique diagnostic and therapeutic challenges due to its distinct histological features, indolent clinical course, and potential for aggressive local invasion. ACC occurs at a rate of less than 0.2/100 000 individuals annually[3]. It is an epithelial cell-derived malignancy characterized by perineural invasion, slow growth, and a high propensity for local recurrence even after seemingly successful treatment. ACC of the trachea often presents with cough, stridor and shortness of breath. Diagnosis involves a multidisciplinary approach, including detailed patient history, physical examination, imaging studies and biopsy. Surgery remains the primary treatment. Adjuvant therapy which includes radiotherapy is often recommended to reduce the risk of local recurrence. However, the optimal radiation therapy regimen is still being explored.

The prognosis of ACC of the trachea depends on various factors, including tumor stage, histological grade, and response to treatment. Despite its indolent nature, ACC can be locally aggressive and may recur even after apparently successful resection. A multidisciplinary approach is crucial in managing ACC of the trachea to ensure optimal diagnosis, treatment planning, and follow-up care. Through a comprehensive understanding of its epidemiology, pathogenesis, clinical manifestations, diagnosis, treatment options, and prognosis, clinicians can enhance awareness, facilitate early diagnosis, and optimize management strategies for patients affected by this rare disease.

CASE PRESENTATION:

A 49-year-old woman, experiencing worsening dyspnea and cough resistant to standard treatments for 6 months, was examined. On January 31, 2017, a flexible bronchoscopy revealed an obstructive tumor in the upper trachea, which a biopsy confirmed as adenoid cystic carcinoma. To assess the disease's scope and exclude distant metastases, computed tomography (CT) scans and additional imaging tests were performed.

Examination of the left lower chest showed reduced breath sounds. Routine blood and urine analyses conducted during laboratory tests showed no irregularities (Figs 1 and 2).

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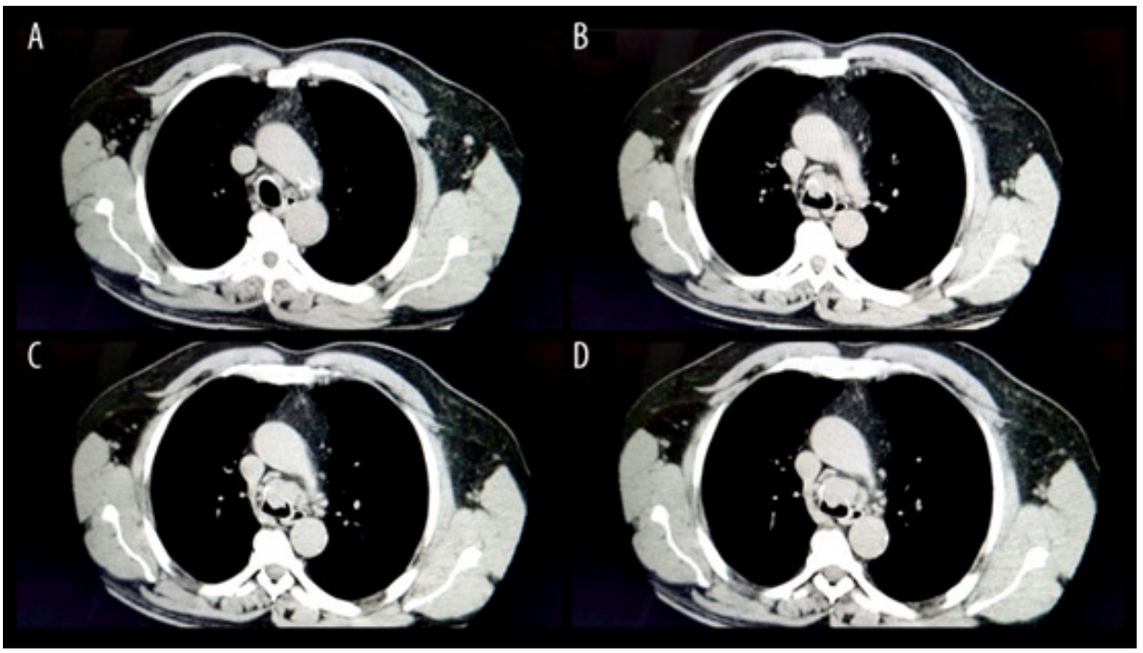

Fig. 1: Computed tomography scan of the chest revealed the lesion in the trachea.

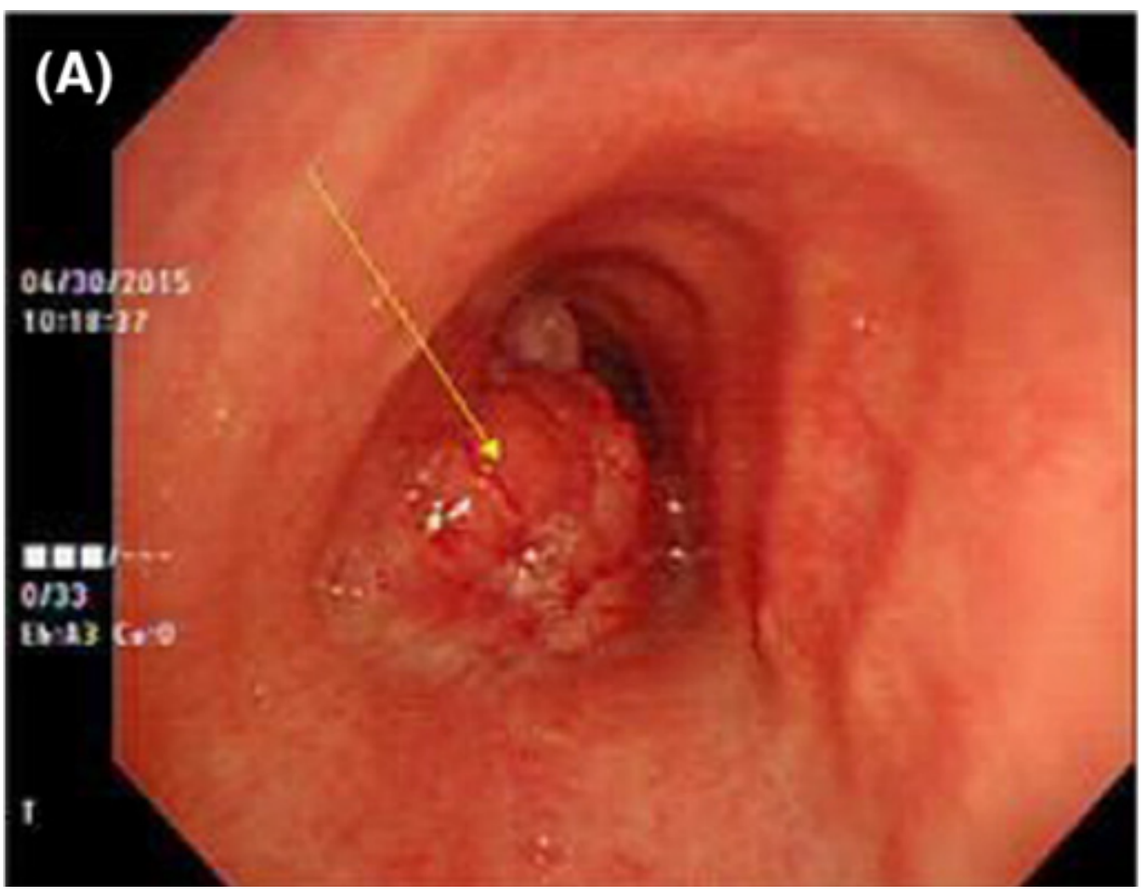

Fig. 2: Bronchoscopic examination before treatment showed a large polypoid mass inside the lumen, originating just before the carina and nearly completely obstructing the lumen.

Diagnosis

Histopathological examination of the biopsy specimen revealed characteristic cribriform and tubular patterns, consistent with adenoid cystic carcinoma. Immunohistochemical staining demonstrated positivity for cytokeratin CK7, negative for CK20 and S-100 protein, confirming the diagnosis. Clinically staged as stage I T1 N0 M0 with tumor about 2 cm (Fig. 3).

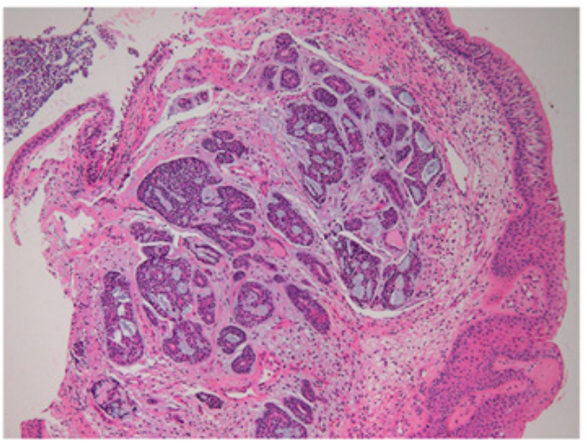

Fig. 3: Histopathological analysis revealed a predominance of the cribriform type within a heterogeneous growth pattern, identifying it as adenoid cystic carcinoma.

Treatment and outcome

A multidisciplinary team of thoracic surgeons, radiation oncologists, and medical oncologists carefully evaluated the patient's case to determine the optimal treatment plan. Given the location and characteristics of the tumor, surgical resection was deemed the most appropriate option.

On February 5, 2017, the patient underwent surgical resection, which involved tracheal sleeve resection and primary anastomosis. Histopathological examination showed an adenoid cystic carcinoma, Grade II (according to the Van Weert 2015 grading system), with a tumor size of 20 mm in its greatest dimension. Perineural invasion was identified, but no definitive lymphovascular invasion was noted. Notably, the tumor extended to the surface epithelium with focal ulceration, and tumor cells were present at the proximal, distal, and inked circumferential margins. One lymph node dissected from the right phrenic nerve tested negative for metastases (0/1) and one lymph node from the Station 4 paratracheal lymph node tested negative for metastasis (0/1). A subsequent biopsy from the new distal margin confirmed the presence of adenoid cystic carcinoma. Dissection of one paratracheal lymph node at station 4 showed no evidence of metastases (0/1).

Following surgery, the patient received adjuvant radiation therapy using volumetric modulated arc therapy and (megavoltage) radiography for a total dose of 6600 cGy in 33 fractions to the target volume. Treatment was administered five times per week from March 19 to May 10, 2017, with a dose of 2.0 Gy per fraction (Figs. 4–7).

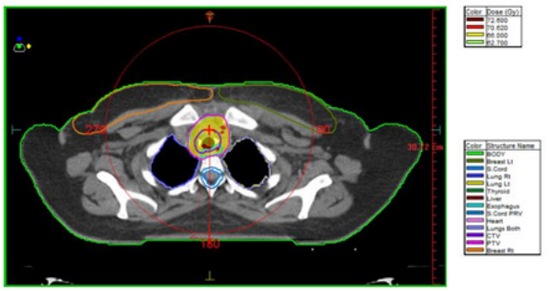

Fig. 4: Shows a three dimensional reconstructed AXIAL computed tomography scan of the chest, depicting the radiotherapy plan.

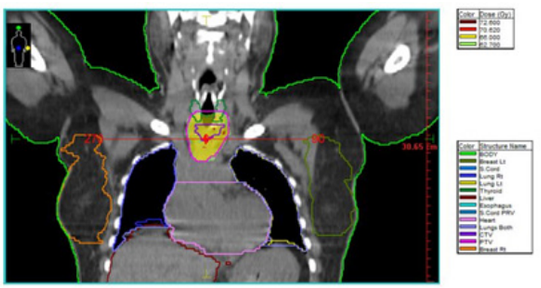

Fig. 5: Shows a three dimensional reconstructed SAGITAL computed tomography scan of the chest, depicting the radiotherapy plan.

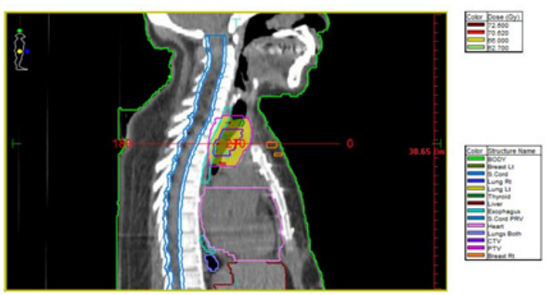

Fig. 6: Shows a three dimensional reconstructed CORONAL computed tomography scan of the chest, depicting the radiotherapy plan.

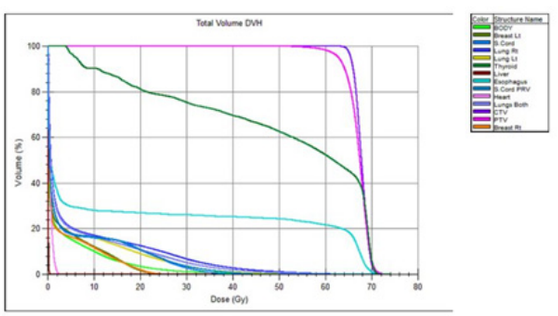

Fig. 7: DVH (dose volume histogram).

DISCUSSION

Primary tracheal cancer is an exceptionally rare condition, occurring at a rate of about 0.1/100 000 individuals each year[4,5]. Tracheal ACC grows slowly, has a propensity for spreading along nerves, and often recurs locally even after being completely removed. To achieve the best possible outcomes, a comprehensive treatment strategy involving multiple specialties recommends adjuvant radiation therapy following surgical removal when no cancerous margins are found.

Follow-up is essential given the risk of distant metastases and the potential for recurrence years later. There are both malignant and benign forms of tracheal tumors, with the former being more common in adults[6]. In our case, the tumor was in the proximal part of the trachea. Similar to what our patient experienced, the symptoms of tracheal ACC can vary, but dyspnea, or shortness of breath, is usually the primary complaint. This symptom is often chronic, worsening gradually over time. Other possible symptoms include coughing, hemoptysis, stridor, or frequent respiratory infections, all of which depend on the tumor's size and location[7,8].

Identifying the optimal treatment approach for tracheal ACC requires an accurate diagnosis[9,10]. CT and MRI are extensively utilized for diagnostic assessments, helping to precisely define the tumor's location, size, and the extent of its spread within the trachea. Nevertheless, to conclusively confirm the diagnosis and obtain tissue for histological analysis, a bronchoscopy with biopsy is indispensable. Bronchoscopy allows for direct visualization of the tracheal lesion and facilitates the collection of tissue samples for biopsy purposes.

When complete removal of the tumor is not possible, palliative treatments such as tracheal stenting or debulking surgery often considered to improve symptoms[11]. For those diagnosed with ACC, radiation therapy is commonly recommended due to the tumor's propensity for localized recurrences. Postoperative radiotherapy is suggested for patients with positive lymph nodes or positive surgical margins[12,13]. Moreover, some retrospective studies have shown a correlation between improved disease-free survival rates following surgery and enhanced local control of the disease[14].

Despite improvements in controlling the disease locally, several studies have not observed an improvement in overall survival[15–17]. The optimal radiation therapy dose and regimen for tracheal ACC is still not well-defined. Treatment decisions should be personalized, considering patient-specific factors, the extent of surgical removal, and whether surgical margins are clear. Postsurgery, a radiation dose of about 60 Gy is typically advised for the original tumor site if complete resection (R0) was achieved[18,19].

We provided adjuvant radiation using volumetric modulated arc therapy (VMAT) and 6-megavoltage (MV) radiography, targeting the initial tumor area plus a safety margin. The treatment was planned to give a total of 6600 cGy across 33 fractions, focusing on the CTV and extending an additional 1 cm to form the planning target volume (PTV), which includes the postoperative bed margins. To minimize the chance of local recurrence, the therapy was given 5 days a week, with 2.0 Gy per session from March 19 to May 10, 2017. Over a five-year monitoring period, follow-up imaging showed no signs of distant metastases or recurrence.

The CT scan conducted on May 13, 2023, revealed several surgical staples in the lower trachea but no growing or persistent lesions. This suggests that the aerodigestive tract remains normal. The parotid, submandibular, and thyroid glands appeared unremarkable. Major neck vessels were found to be normal, and no significant osseous lesions were detected, indicating absence of persistent or recurrent disease.

CONCLUSION

Tracheal ACC is a rare and challenging malignancy. Prompt recognition and appropriate management are essential to achieve optimal outcomes. This case report describes the uncommon occurrence of proximal tracheal ACC and demonstrates the effectiveness of individualized radiation therapy in combination with surgical resection, resulting in a 5-year survival without recurrence. This case underscores the importance of a multidisciplinary approach in the management of tracheal ACC. By tailoring treatment plans to the specific characteristics of each patient, we can improve outcomes and enhance the quality of life for those affected by this rare disease.

CONFLICT OF INTEREST

There are no conflicts of interest.

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