

Adenoid Cystic Carcinoma of the Trachea: Case Report and Literature Review

Maroua Benlemlih^{1*}, El Amin Marnouche¹, Mouhcine Hommadi¹, Abdelhak Maghous¹, Amine Bazine¹, Kessab Amine², Massine El Hammoumi³, Oukabli Mohamed², Khalid Andaloussi¹, El Hassane Kabiri³, Mohammed Elmarjany¹, Khalid Hadadi¹, Hassan Sifat¹ and Hamid Mansouri¹

¹Department of Radiotherapy, Mohammed V Military Teaching Hospital, Mohammed V University in Rabat, Morocco

²Department of Pathology, Mohammed V Military Teaching Hospital, Mohammed V University in Rabat, Rabat, Morocco

³Department of Thoracic Surgery, Mohammed V Military Teaching Hospital, Mohammed V University in Rabat, Rabat, Morocco

Abstract

Tracheal cancer is a rare neoplasm that accounts for only about 0.1% of all respiratory tract tumors. Cystic adenoid carcinoma (or cylindroma) represents the second histological type of tracheal tumors in order of frequency. These lesions are characterized by slow local growth, perineural invasion, and potential local and distant recurrence. Complete surgical resection offers the patient a better opportunity of prolonged survival or complete remission. The addition of photon- or electron-beam radiation seems to influence local control but does not affect survival. In this paper, we report a clinical case describing the characteristics and management of adenoid cystic carcinoma of the trachea in 59 years-old men.

Keywords: Adenoid cystic carcinoma • Trachea • Radiotherapy • Cancer

Abbreviations: ACC: Adenoid Cystic Carcinoma; RT: Radiotherapy; CTV: Clinical Target Volume; PTV: Planning Target Volume; SCC: Squamous Cell Carcinoma; CT: Computed Tomography

Introduction

Adenoid Cystic Carcinoma (ACC) is a rare primary malignancy of trachea. The clinical and pathologic features of ACC of the trachea were initially reported in 1859 by Bill Roth [1]. It represents 0.09% to 0.2% of all thoracic tumors [2, 3]. ACC is characterized by slow growth, and distant metastasis has been reported as late as 25 years after diagnosis [4, 5]. Currently, because of their locally invasive and potentially distant evolution in advanced forms, they are classified as malignant tumors. The clinical presentation of these tracheal localization lesions is often stereotyped. Inspiratory dyspnea is often in the foreground. Surgery is considered as the treatment of choice for ACC, and most articles have focused on the surgical outcome. Radiotherapy (RT) was traditionally used as an adjuvant treatment for controlling microscopic disease or as a salvage treatment for unresectable disease; however, the exact role of RT remains unclear due to the rarity of such reports. We present here a clinical case of ACC of the lower trachea and different treatment modalities [6].

Case Report

Our patient is 59 years old men with a history of tobacco use at a rate of a pack a day over 30 years. The symptoms started 2 years ago with dyspnea and dry cough treated in pulmonology as chronic obstructive pulmonary disease. But no improvement was noted.

A chest scan was done and showed tracheal thickening at T10 height with an endoluminal tumor reducing tracheal lumen measuring 22 × 25 × 35 mm without any mediastinal adenopathy (Figure 1). A bronchoscopy revealed a

tracheal tumor obstructing of 85% of the tracheal lumen extending of 35 mm. a biopsy was performed. Histopathological examination found a myoepithelial carcinoma of trachea. No distant metastasis was found after extension assessment.

The patient underwent surgery with tumor resection and termino-terminal tracheal anastomosis without mediastinal lymph node dissection. The surgical approach was right posterolateral tracheotomy, and the tumor was palpable with well encapsulated extra luminal buds.

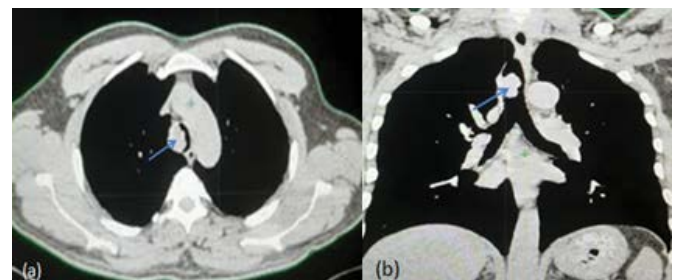


Figure 1. Chest scannography with axial (a) and coronal (b) views showing a tracheal endoluminal tumor (blue arrow).

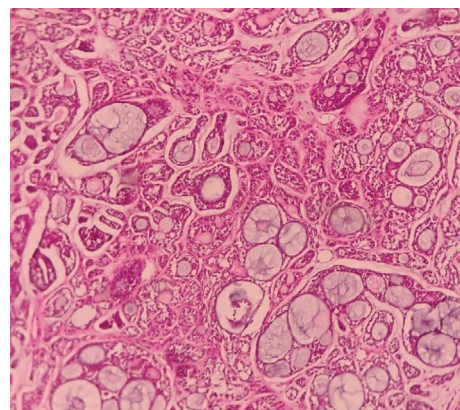


Figure 2. Hematoxylin and eosin (HE) stain revealed adenoid cystic carcinoma.

***Address for Correspondence:** Maroua Benlemlih, Department of Radiotherapy, Mohammed V Military Teaching Hospital, Mohammed V University in Rabat, Rabat, Morocco; Tel: +212674910122; E-mail: maroua.benlemlih@gmail.com

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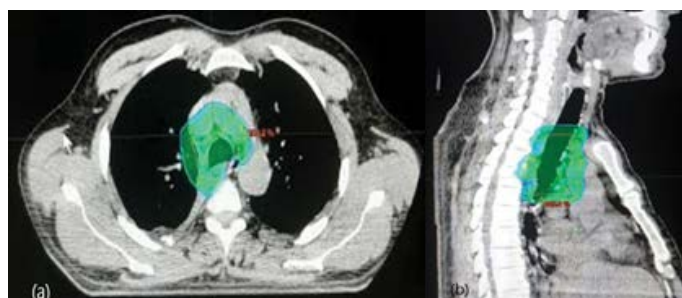


Figure 3. Axial (a) and coronal (b) scan views of radiotherapy planning plan showing dose distribution (95% isodose).

Histological and immune histochemical findings showed an adenoid cystic carcinoma of the trachea (Figure 2). The surgical margins were negative.

Adjuvant radiotherapy at a dose of 54 gray was delivered for 27 fractions with intensity-modulated radiotherapy. The Clinical Target Volume (CTV) expanded in 3 cm longitudinal from the tumor bed and the Planning Target Volume (PTV) was expanded 1 cm in both longitudinal and axial plans from the CTV (Figure 3).

The follow up of 2 years with regular bronchoscopy and chest scan didn't show any local or distant recurrence.

Discussion

Adenoid cystic carcinoma originally arises from the salivary glands and is a slowly progressing malignancy, with late metastasis to the lung, bone, and brain [7]. The most common site in the airway is the trachea and other localizations such as the glands of the bronchial mucosa have been described [8]. ACC spreads most commonly by direct extension, submucosal or perineural invasion, or hematogenous metastasis. More than 50% of patients with tracheal ACC have hematogenous metastases. Pulmonary metastases are the most common and can remain asymptomatic for many years [9]. Lymphatic spread is uncommon in patients with tracheal ACC.

In contrast to tracheal Squamous Cell Carcinoma (SCC) that occur in men approximately 90% of the time, primary tracheal ACC is found in men and women with almost equal frequency. The ages reported for tracheal ACC ranged from 45 to 60 years without any tobacco imputation [10]. Our patient was 59 years old.

Patients with ACC usually present with symptoms such as coughing, wheezing and dyspnea and are often treated for asthma for months to years before being correctly diagnosed. Patients with tracheal malignancies demonstrated the most frequent symptoms such as wheezing or stridor, dyspnea, hemoptysis, and coughing. Few patients presented with hoarseness and weight loss [11]. This clinical symptomatology is long tolerated because of the large size of the trachea and the slow growth of the tumor. The average time to revelation after the first symptom is often greater than six months as showed in our experience since the patient spent 2 years treated as chronic obstructive pulmonary disease before discovering the real etiology.

The chest x-ray may appear normal or have lateral endotracheal opacity or endotracheal tumor. Thoracic Computed Tomography (CT) is necessary to evaluate the peri-tracheal extension of the tumor and to discover any nodes or secondary pulmonary lesion. Then, a tracheobronchial endoscopy is mandatory to localize exactly the tumor, describe the macroscopic appearance and realize a biopsy for histologic confirmation of diagnosis.

Treatment options include surgery alone, radiation therapy alone, or a combination while chemotherapy has no place outside metastatic forms [12]. Surgery is the mainstay of treatment and consists of tumor resection and end-to-end anastomosis with or without reconstruction. However, the extent of the resection makes the anastomosis more difficult, causing complications. Lymph nodes dissection should not be too extensive not to compromise tracheal vascularization. In previous studies, the operative mortality rate was an

average of 12% (range 5%-14%) [13]. Reported complications include trachea esophageal fistula, pharyngeal or esophageal leak, anastomotic separation, wound dehiscence, vocal cord paralysis, temporary tracheotomy, dysphagia, ileus, and pneumonia [14]. In our patient, thoracic surgeons realized a cuneiform tracheal resection removing the tumor implantation base followed by lateral tracheal anastomosis but the desaturation didn't allow lymph nodes dissection. Fortunately, no post-operative complication was noted.

Radiation therapy for ACC of salivary glands has been found to provide improved local control of tumors but did not affect survival [15]. This treatment has not been investigated for tracheal ACC. The role of post-operative adjuvant radiotherapy remains uncertain due to the rarity of such reports. It is reasonable to assume that adjuvant radiation therapy may be beneficial and likely delays or reduces the incidence of local recurrence in the airway without improving overall survival as showed in this large cohort of 39 ACC of trachea treated with adjuvant or definitive radiotherapy [7]. So, with limitations of randomized comparison, it still seems reasonable to recommend adjuvant radiotherapy for all patients undergoing resection, and certainly for those in whom the final pathologic examination identifies residual tumors at the resection margins. Therefore, there is a place for radiotherapy as definitive treatment for unresectable tumors. Adjuvant radiation was performed after a tracheobronchial fibroscopy of control at 8 weeks of surgery in our institution at a dose of 54 gray considering the clear resection margins. To note that the main complication of post-operative radiotherapy reported in the literature is tracheal stenosis, so the risk of tracheal stenosis should be carefully considered when determining the radiotherapy plan or dose.

Indeed, ACC is a gradual low malignancy tumor usually associated with prolonged survival. However, when it is associated with distant metastases survival is frequently less than 2 years. Complete surgical resection offers the patient a better opportunity of prolonged survival or complete remission. After a follow up of 2 years, our patient is still stable with good control.

Conclusion

ACC is a rare primary tracheal malignancy. Hemoptysis, nonproductive cough, dyspnea, chest pain and weight loss are the common initial symptoms. The time from first symptoms to diagnosis varied, ranging from weeks to more than 1 year. This disease is commonly misdiagnosed as asthma. Complete surgical resection provides the patient with the best chance of prolonged survival or even complete remission. Post-operative radiotherapy may have some effect on local control but did not affect survival rate.

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