Adenoid Cystic Carcinoma of the Trachea: A Case Report

Mohamed Ait Erraisse, Sa El Mazghi, Touria Bouhafa and Khalid Hassouni

Hospital Center University Hassan II, Fes, Morocco

*Corresponding author: Mohamed Ait Erraisse, Hospital Center University Hassan II, Fes, Morocco, Tel: +212619948046; E-mail: aiterraissemohamed@gmail.com

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Abstract

Primary malignant tumors of the trachea are rare. It represents 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. We report here a clinical case detailing the management of adenoid cystic carcinoma of the trachea in a 17-year-old boy.

Keywords: Trachea; Cylindroma; Carcinoma

Introduction

Cystic adenoid carcinomas (CAK) are rare primary salivary gland tumors representing 0.09 to 0.2% of all thoracic tumors [1,2]. These tumors, first described by Billroth in 1856 [3], were formerly considered to be tumors of intermediate malignancy. Currently, because of their locally invasive and potentially distant evolution in advanced forms [4], they are classified as malignant tumors [5]. The clinical presentation of these tracheal localization lesions is often stereotyped. Inspiratory dyspnea is often in the foreground. The treatment modalities are multiple but surgery remains the treatment of choice. We present here a clinical case of CAK of the upper trachea.

Case Report

Our patient is a 17-year-old boy with no pathological history. The symptoms started 9 months ago with shortness of breath and cough followed by dysphagia and notion of false roads without hemoptysis.

A bronchoscopy was able to show a tracheal tumor obstructing about 80% of the tracheal lumen located 2.5 cm from the vocal cords extended on 2.8 cm (Figure 1).

A biopsy was performed after showing the pathological examination of adenoid cystic carcinoma. Cervico-thoracoabdominal CT showed tracheal thickening at T2 height with an endoluminal tumor reducing tracheal lumen without distant metastasis. The patient underwent surgery with tumor resection and termino-terminal tracheal anastomosis and mediastinal lymph node dissection.

On histopathological examination: histological and immunohistochemical findings showed an adenoid cystic carcinoma of the trachea, 2.5 cm long axis. The upper margin was negative. The lower resection margin was microscopically tumoral (R1) since surgical resection was at maximum possible (tracheal length 6 cm). Lymph node dissection was negative in total (17 negative ganglia): pre-tracheal, lateral tracheal, right jugulo-carotid, left recurrence. Subsequently, the patient received adjuvant radiotherapy on the tumor bed at a dose of 66 Gy given the positive margin.

Figure 1: Endoscopic view of the tumoral tracheal stenosis.

Figure 2: Endoscopic view of the inflammatory stenosis.
During follow-up, he had tracheal stenoses on the anastomosis that were dilated three times with two-time silicone prosthesis and a biopsy of inflammatory stenosis with no signs of malignancy (Figure 2). During the last episode, the patient had acute dyspnea requiring hospitalization in intensive care unit with attempted dilation but without success. Patient died after 18 months of follow up.

Discussion

Cystic adenoid carcinomas, formerly called cylindromas, are malignant epithelial tumors that develop most often from the main and accessory salivary glands. Other localizations such as the glands of the bronchial mucosa have been described [1]. These tumors are mostly observed at a younger age than other cancers, with no predominance of sex or tobacco imputation. Cystic adenoid carcinomas are often of late diagnosis, indeed the clinical symptomatology is often very moderate and misleading, evoking more asthma, the main manifestation being dyspnea. 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 [1,2].

The chest x-ray may appear normal or have lateral endotracheal opacity or endotracheal tumor. Thoracic computed tomography (CT) is used to evaluate the peri-tracheal extension of the tumor and to discover any nodes or secondary pulmonary lesion. The tracheobronchial fibroscopy is essential, it allows to specify the seat and confirm the diagnosis by performing a biopsy often difficult to read [2]. The treatment is based on three therapeutic modalities, namely surgery, radiotherapy and interventional endoscopy. Chemotherapy has no place outside metastatic forms [3,4]. Surgery consists of resection of the tumor with a tracheal cut and an end-to-end anastomosis, followed by dissection of the lymph nodes. The extent of the resection makes the anastomosis more difficult, causing complications or postoperative recurrence. Lymph node dissection should not be too extensive not to compromise tracheal vascularization. The periooperative mortality rate varies between 9 and 13% depending on the series [3,4]. radiotherapy is indicated when resection is incomplete or impossible or after local recurrence and lymph node involvement. In the case of a severe tumor obstruction, the endoscopic laser resection quickly allows a disobstruction, which will eventually be followed by an optimal local treatment [5]. Endobronchial brachytherapy can be used to increase the total dose of irradiation and improve the rate of local control [6].

Conclusion

Adenoid cystic carcinoma of the trachea is a rare tumor. Its diagnosis and surveillance is based on bronchoscopy associated with biopsy. Computed tomography better assesses the extent and peri-tracheal extension of this tumor. Therapeutic management is essentially based on surgery and radiotherapy. Post-therapeutic stenosis remains the main complication in the absence of reconstruction.

References