

Adenoid Cystic Carcinoma of the Trachea

Carcinoma adenoide quístico de tráquea

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CASE REPORT

This report describes the case of a 51-year-old female, non-smoker, without any relevant medical or surgical history, who is an information technology professor. She was referred to pulmonology consultation due to an expiratory stridor of two months of evolution that was partially interfering with her work, under suspicion of possible bronchial asthma. The physical examination only revealed said expiratory stridor. The forced spirometry showed a flow-volume curve suggestive of irreversible airway obstruction with the following values: forced vital capacity (FVC) 95%, forced expiratory volume on the first second (FEV₁) 52.8% and FEV₁/FVC ratio 47.23%, with negative bronchodilator test. Basing on such findings, hospitalization was indicated in order to study a possible intrathoracic mass. The following imaging tests were done: computed tomography (CT) and positron emission tomography (PET-CT), showing a large mediastinal mass (Figures 1 and 2).



Figure 1. Sagittal section of chest CT showing nodular lesion of retrotracheal and right paraesophageal location, with a length of 9 cm, 3 cm on the anteroposterior axis and 3.3 cm on its transverse axis. Its caudal end is shown (red arrow).

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Figure 2. Sagittal section of PET-CT showing the mass with increased uptake of 18-fluorodeoxyglucose (18 FDG) and retrotracheal location in posterior mediastinum, suggestive of malignant neoplasm.

Also, a flexible bronchoscopy was performed (Figure 3), showing great damage to the tracheal *pars membranosa*. No samples were taken due to risk of bleeding. It was decided to do video-assisted thoracoscopic surgery (VATS) to obtain histological material from an area that could ensure more safety and control in case of hemorrhage.

Finally, the diagnosis obtained was locally advanced adenoid cystic carcinoma of the trachea. Chemotherapy with carboplatin and paclitaxel was indicated and completed in 3 cycles plus radiotherapy fractionated in 33 sessions. The clinical tolerance of the patient to this treatment was adequate, and she didn't develop any notable complications. Approximately 5 months after finishing treatment, there was certain mass size reduction, and it remained stable during successive radiological controls until now, with a length of 6 cm, anteroposterior diameter of 2.4 cm and transverse diameter of 2.1 cm. Also a reduction in the 18-FDG uptake could be seen in the subsequent control PET-CT. At present, the patient is under close follow-up for medical oncology and radiation oncology, and the possibility of resection has been discarded.

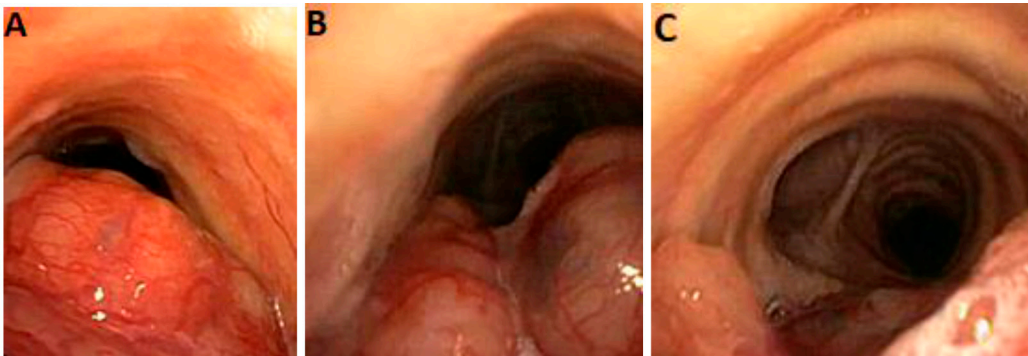


Figure 3. Flexible bronchoscopy showing a largely vascularized tumor of hard consistency that invades the *pars membranosa* and extends from the subglottic region (A), affects the middle third (B) and reaches the distal third of the trachea (C).

DISCUSSION

The adenoid cystic carcinoma of the trachea is an exceptional clinical condition; with an incidence of 0.1-0.2 cases every 100,000 inhabitants per year, it represents the second most common primary tracheal malignant neoplasm following the squamous cell carcinoma^{1, 2}. It poses a diagnostic challenge, due to its nonspecific, insidious symptoms. The typical patient is between 50-70 years old, no sex preference, non-smoker, and shows progressive dyspnea in most cases. In its differential diagnosis, it is important to consider the tracheal squamous cell carcinoma, more common in smokers and typically occurring in association with hemoptysis.

At present there isn't any agreed standard for obtaining a confirmation diagnosis and subsequent staging³. However, there seems to be agreement among the specialized centers that the length of tracheal damage is the variable that defines tumor resection. For lesions larger than 5 cm, like the one evidenced in this report, surgical treatment is not recommended¹. Radiation therapy associated with chemotherapy is recommended in all cases, especially if there is extracapsular extension, perineural, bronchial or vascular invasion or associated adenopathies. 5-year overall survival in resectable cases is between 50-80%, and drops to 30% in the rest of the patients^{1, 4, 5}.

Conflicts of interests: The authors declare no conflict of interest in relation to the contents of this article.

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