

Tracheal cystic adenoid carcinoma: Case report presentation and literature review

Carcinoma adenoide quístico traqueal: presentación de caso clínico y revisión de literatura

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ABSTRACT

Introduction: The adenoid cystic carcinoma of the airway is a rare tumor that originates from the submucosal glands of the tracheobronchial tree. Due to the usual delay that occurs between symptoms and diagnosis, and the propensity of this tumor to expand through the perineural pathways and submucosa, the recommended treatment is surgical resection with postoperative radiation therapy. Survival is determined by the presence of distant metastasis.

Case report: 70 year-old female patient with a history of arterial hypertension, COPD (former smoker, 34 pack/years) who came to the Emergency Service with episodes of hemoptysis and previous dyspnea with a score of 3-4 according to the mMRC (modified Medical Research Council) dyspnea scale.

Discussion: Malignant neoplasms of the trachea are very rare, and data related to them is limited. The most important prognostic factors in primary malignant tumors of the trachea are: early diagnosis, cancer staging, histology, and treatment options.

Conclusions: Early detection may be associated with increased resectability rates and even prolonged survival.

Key words: Hemoptysis; Adenoid cystic carcinoma; Bronchoscopy; Surgery; Radiation therapy

RESUMEN

Introducción: El carcinoma adenoide quístico de la vía aérea es un tumor poco común, que se origina de las glándulas submucosas del árbol traqueobronquial. Por el usual retraso entre los síntomas y el diagnóstico, y por la propensión de este tumor para expandirse a través de los haces perineurales y submucosa, el tratamiento recomendado es la resección quirúrgica con radioterapia posoperatoria. La supervivencia está determinada por la presencia de metástasis a distancia.

Caso clínico: Paciente de sexo femenino de 70 años de edad con antecedentes de hipertensión arterial, EPOC (exabajaquista 34 paquetes/año) que acude a servicio de urgencias con episodios de hemoptisis y disnea mMRC 3-4 previa.

Discusión: Las neoplasias malignas de la tráquea son muy raras y los datos relacionados con ellos son limitados. Los más importantes factores pronósticos en las enfermedades primarias malignas de la tráquea constituyen el diagnóstico temprano, estadiaje del tumor, histología y opciones de tratamiento.

Conclusiones: La detección temprana puede estar asociada con el incremento de las tasas de resecabilidad e, incluso, supervivencia prolongada.

Palabras clave: Hemoptisis; Carcinoma adenoide quístico; Broncoscopia; Cirugía; Radioterapia

INTRODUCTION

Primary tracheal tumors account for less than 1%¹ (0.1% to 0.4%) of all malignant respiratory diseases.² They are typically malignant in adults (80-90%) and benign in children (60-70%).³

A primary tracheal carcinoma is a malignant tumor located between the first tracheal ring and the carina.⁴ Data related to these tumors are limited due to the small number of cases reported in publications, posing a diagnostic and therapeutic challenge.⁵

Squamous cell carcinoma comprises two-thirds of primary tracheal tumors in adults. It usually presents around the age of 60 and predominantly affects men. Adenoid cystic carcinoma is the second most common malignant tracheal tumor after squamous cell carcinoma.⁶ It is typically found in young patients, occurring between the fourth and fifth decades of life, and affects both men and women equally.⁷

Adenoid cystic carcinoma, also known as cylindroma, was first reported by Billroth in 1856. It is a rare malignant tumor of the head and neck, accounting for approximately 10% of tumors localized in this region. It originates in the salivary glands, most commonly in the parotid gland.⁸ Rarely, it can occur in the trachea⁹ originating in the submucosal glands of the tracheobronchial tree. Depending on its location, it can be classified as laryngeal (including subglottis) or tracheobronchial. Laryngeal adenoid cystic carcinoma is extremely rare, with approximately 40 cases reported in the last 41 years. Most adenoid cystic carcinomas develop centrally in the trachea (64.6%) and main bronchi (19.5%).

The primary presenting symptom is often dyspnea. Definitive diagnosis is delayed, and most cases are diagnosed when the disease is already at an advanced stage.

Recent studies, such as the one by Hämetoja et al, have shown that the JC polyomavirus (JCPyV) can be found in samples from minor salivary glands and detected through quantitative polymerase chain reaction (qPCR). However, the prevalence of JCPyV positivity in adenoid cystic carcinoma was low, and the viral copies detected were insufficient to establish its role in the carcinogenesis of this tumor.¹⁰ Other studies suggest a potential role of the human papillomavirus (HPV) in the carcinogenesis of adenoid cystic carcinoma.

Selection criteria and treatment indications for this condition are not consistent. Even patients with resectable disease are often managed with palliative treatment, likely due to the lack of available prospective studies that evaluate and compare treatments, which are almost impossible to conduct given the rarity of these tumors.

The choice of treatment modality clearly affects survival. Surgery has been shown to be superior to radiation therapy in many studies. The 5-year survival rate varies between 41% and 57% with surgical management. In patients treated with radiation, the 5-year survival rate varies from 6% to 11%. For this reason, surgery should be considered in most cases, including those of advanced disease.

The delicate arterial and lymphatic network could explain the rarity of hematogenous metastases and the relative frequency of regional lymph node metastases at initial presentation. Late metastases can occur more commonly in the lungs, but this condition can also spread to the brain, bones, liver, thyroid, and spleen. Adenoid cystic carcinoma is also known for its tendency to cause neutropenia and for recurring locally or regionally many years after initial presentation and treatment. The relatively low incidence of adenoid cystic carcinoma in the periphery of the lung is likely associated with the distribution of glandular cells. In a review of 15 cases, Moukarbel et al reported a local recurrence rate of 33% and a distant metastasis rate of 67%, primarily to the lungs.

Epidemiology

In Argentina, in the year 2017, cancer-related mortality was reported at 118 and 87 deaths per every hundred thousand males and females, respectively. Lung cancer determined the highest number of deaths among malignant tumors in 2017, with 9,485 deaths, accounting for 15% of all cancer-related deaths and 20% of deaths from this cause in males.¹¹

In the United States, adenoid cystic carcinoma represents 2 cases per one million people annually. Other data report 2 to 6 new cases per million people each year, accounting for less than 0.1% of cancer deaths per year.¹²

Due to the rarity of the adenoid cystic carcinoma of the airway, prospective studies that assess prognostic factors, treatment, and outcomes are not feasible and do not allow for external validity

determination. Therefore, cases from institutional series serve as an important guide for the therapeutic approach.

Cigarette smoking history is commonly associated with squamous cell carcinoma of the trachea; however, there are no specific risk factors associated with adenoid cystic carcinoma.

Pathology

Primary tracheal tumors can originate from the respiratory epithelium (squamous cell carcinoma, adenocarcinoma), the salivary glands (adenoid cystic carcinoma, mucoepidermoid carcinoma), and mesenchymal structures (sarcoma, lymphoma).

Adenoid cystic carcinoma is a distinct type of carcinoma that arises from both major and minor salivary glands. Less commonly, it can originate in the seromucinous glands of the upper and lower respiratory tract, which have been shown to decrease from the supraglottis to the glottis, subglottis, and trachea.¹³

According to the World Health Organization classification, adenoid cystic carcinoma is defined as a basaloid tumor composed of epithelial and myoepithelial cells with various morphological configurations, including tubular, cribriform, and solid patterns.¹⁴ The cribriform pattern is the most common, characterized by uniform cells arranged in nests separated by cystic spaces containing mucinous material.

Histologically, these tumors consist of two main cell types: ductal (luminal) and myoepithelial (ab-luminal) cells.

Macroscopically, adenoid cystic carcinoma exhibits exophytic nodular growth, leading to stenosis of the tracheal lumen. It has a propensity to spread along the submucosal and perineural planes, with only 10% of patients developing lymph node or distant metastases. The presence of perineural invasion is a distinctive feature of the tumor and is associated with a tendency to develop recurrent disease after surgical resection, likely due to the higher likelihood of microscopic residual disease at the resection margins or beyond and its strong propensity to invade the nerves.¹⁵

The pathological distribution is related to the prognosis. Adenoid cystic carcinoma has shown greater survival, compared to squamous cell carcinoma

Mitsuaki et al described an abrupt transformation of a low-grade or well-differentiated tumor within a tumor with a high-grade component without a spectrum of the original tumor; this is called undifferentiated adenoid cystic carcinoma of the trachea, which is an extremely rare and highly aggressive tumor.¹⁶

Clinical presentation

Tracheal tumors can often go undiagnosed for months or even years due to their slow growth and silent nature, and are typically discovered at a later stage. In a study including 52 patients between March 1995 and March 2012, Chen et al described that the average duration of symptoms before diagnosis was 18 months, with a range from 1 to 98 months.

Many patients present with symptoms such as dyspnea, wheezing, and chronic cough, which are often confused with conditions like asthma. In smokers, these symptoms can be mistaken for chronic obstructive pulmonary disease or chronic bronchitis, because these tumors do not cause symptoms until they occlude more than 50% of the tracheal lumen diameter. Exertional dyspnea does not typically develop until the trachea has reduced its lumen to less than 8 mm, and once the lumen becomes less than 5 mm or 75% of its original size, dyspnea can also occur at rest.¹⁷ This tumor can progress to the point of compromising the airway and causing fatal secondary asphyxia, as reported by Huston et al.

In a study involving 82 patients at the Shanghai Chest Hospital in China from March 2001 to April 2012, Zhao et al described that the primary symptom of tracheal adenoid carcinoma was dyspnea (66%), followed by cough (13.2%), hemoptysis (13.2%), and stridor (3.8%).

Irritation or ulceration of the mucosa can lead to cough and hemoptysis, while the invasion of adjacent structures can result in dysphagia. Distant metastases occur in less than 10% of patients.

Hemoptysis is the main symptom in patients with squamous cell carcinoma, and typically leads to early diagnosis within 4 to 6 months. Adenoid cystic carcinoma presents with wheezing or stridor as the primary symptom, and less than 25% of patients experience early hemoptysis in its course, which explains why symptoms can last for approximately 18 months before a definitive diagnosis is made.

Diagnosis

Patients with symptoms such as shortness of breath and wheezing that do not respond to bronchodilator treatment should prompt us to consider a tracheal tumor among the differential diagnoses.

Lung function tests, for example the spirometry, can reveal fixed upper airway obstruction, evidencing impairment in both inspiratory and expiratory flow-volume curves.

Chest X-rays are rarely diagnostic. The most useful method for assessing the extent and relationship of the tumor with adjacent structures is a CT scan. Imaging studies with multiplanar and three-dimensional reconstruction with internal (virtual bronchoscopy) and external views can demonstrate whether the lesion is inside the lumen, outside the airway, or has characteristics of both.¹⁸ The presence of a soft tissue mass in the trachea with increased uptake of 18-fluorodeoxyglucose (¹⁸F-FDG) on positron emission tomography with multi slice computed tomography (PET/MSCT) is highly suggestive of a malignant tracheal tumor.

The bronchoscopy is a valuable tool for the diagnosis and staging of tracheal tumors because it allows for the collection of tissue samples and the evaluation of the location and extent of the disease, as well as the relationship between the length of the tumor and the trachea. The endoscopic ultrasound can also determine the degree of tracheal invasion.¹³

Bronchoscopic findings may reveal a large mass or circumferential lesion within the trachea. The appearance of the tumor can vary, but it is predominantly red, granular, or fleshy, and easily friable. The borders of the lesion may be poorly defined or diffusely infiltrative. The margins of protruding masses may also show mucosal elevation or vascularity, providing evidence of infiltration beneath the mucosa.

Treatment

Malignant primary tumors are usually treated with surgery, endoscopic resection through various techniques, and radiation therapy. However, only surgery can cure benign tumors and low-grade malignant tumors, achieving long-term survival in tracheal carcinomas. Surgery also provides complete pathological confirmation of the tumor and permanently relieves airway obstruction.

Identifying the extent of local disease is the most important factor in determining the therapeutic management.¹⁹ The decision to resect or irradiate the tracheal tumor will depend on many factors, including the patient's overall health, tumor histology and location, and the length of the airway that could be preserved after resection.

If a patient has life-threatening airway obstruction, resection with rigid bronchoscopy may be used to delay the surgery. However, management with stents or non-adjuvant radiation therapy is not recommended unless the resection cannot be performed.

Surgery

Surgery is the cornerstone of treatment for adenoid cystic carcinoma, and requires a high level of expertise. It is applicable to patients with localized disease and has been associated with a better long-term prognosis.²⁰

Compared to other head and neck cancers, adenoid cystic carcinoma is more challenging in terms of surgical resolution, often resulting in positive margins.

Complete resection is achieved in 42%-57% of cases. It is associated with better survival and is essential due to the high recurrence rate when a residual tumor remains. There is a higher risk of local recurrence and positive surgical margins when the tumor is located in the distal trachea.

Some of the surgical techniques for treating tracheal tumors are: laryngectomy with resection of the upper trachea, larynx, and trachea; tracheal resection; carinal resection without lung resection; and carinal resection with lung resection. Laryngotracheal resection should be preferred over laryngectomy for subglottic tumors. Jiao et al described a new minimally invasive surgical technique in which they performed circumferential tracheal resection and end-to-end anastomosis via thoracoscopy, taking into consideration factors such as tumor size, location, local invasion of the lesion, and the surgeon's experience. This approach was found to be safe, effective, and could serve as a new alternative strategy for treating distal tracheal tumors.²¹

In cases of bronchial localization, some of the surgical techniques include pneumonectomy, carinal resection without lung removal, carinal resection with lung removal, sleeve lobectomy,

and lobectomy. The use of deltopectoral flaps with costal cartilages has been found to be satisfactory.

Absolute contraindications for surgery include the presence of numerous positive lymph nodes, involvement of more than 50% of the trachea, mediastinal invasion of unresectable organs, mediastinum that has received a maximum radiation dose of more than 60 Gy, or previous surgery for distant metastases of squamous cell carcinoma.

For a minority of patients (less than 20%) who present with metastatic disease, resection may be purely palliative, aimed at relieving airway obstruction in cases where a tracheostomy is not feasible.

Bronchoscopy

It is a useful technique for the evaluation and, in several cases, the palliative treatment of the respiratory airways by reducing the tumor or in unresectable patients for stent placement.

Endotracheal tumors can be resected endoscopically for palliation in inoperable patients (e.g., patients with stage T4N3 or higher) or as a means to keep the airway permeable until definitive resection can be performed. Tumors can be removed using biopsy forceps and suction, electrocoagulation, cryotherapy, laser therapy, photodynamic therapy, or argon plasma coagulation. These measures should never be used as curative attempts because they rarely offer long-term survival.

Sato et al described the multi-session endoscopic treatment with argon plasma coagulation in a patient with tracheal adenoid cystic carcinoma. They demonstrated its safety by producing less vapor and smoke, controlling the depth of coagulation (up to 3 to 4 mm maximum), and ensuring safe and effective coagulation, especially in large areas. This method was considered a safe palliative therapy, similar to other methods like electrocautery or Nd-YAG laser for tumor control, with fewer adverse reactions.²²

Endobronchial stents

In patients with unresectable or medically inoperable lesions, reliable and durable palliation can be achieved in 80%-90% of appropriately selected patients through the use of expandable or silicone stents.

Both silicone and self-expanding metallic stents (SEMS) are widely used in case of airway stenosis.

One type of SEMS, known as the AERO stent, combines the characteristics of a metallic and silicone stent covered with a nitinol structure. Its advantages include being insertable via flexible bronchoscopy, easy removal, strong expansion properties, and a lower risk of migration. However, it is associated with a higher risk of infection compared to other stents. Nonetheless, it provides an effective means to improve the patients' quality of life.²³ Himeji et al described two cases of tracheal stenosis secondary to malignant disease in which they used a self-expanding metallic stent (SEMS). They reported no identified complications and improvement in obstructive symptoms, resulting in a clear enhancement of the patients' quality of life.

Radiation therapy

Radiation therapy is indicated as definitive therapy for primary unresectable lesions, in medically inoperable patients, as adjuvant treatment after resection, and for palliation of severe symptoms. Radiation therapy alone has typically been reserved for advanced or unresectable cases. Postoperative radiation therapy should be used in most patients, but post-surgical radiation is also a treatment of choice because surgical margins are often involved.

Negative margins and adjuvant postoperative radiation therapy are associated with improved survival prognosis.

Incomplete resection can be converted to complete resection by administering 60 Gy of postoperative photon radiation therapy, given as five fractions of 2 Gy per week for more than 6 weeks. This treatment eliminates microscopic residual carcinoma in the tumor bed and regional lymph nodes. For macroscopic residual carcinoma, the required doses should be increased to 68-70 Gy, administered as 5 fractions of 2 Gy for more than 7 weeks.

High-dose endobronchial therapy with Iridium-192 has been reported to yield good palliative results with minimal toxicity. However, a small study involving four tracheal neoplasms treated with endobronchial Iridium-192 reported tracheal stenosis in two long-term survivors.

Endotracheal brachytherapy could be a reasonable approach for tracheal carcinomas, as it has shown to improve local tumor control when used after 60-68 Gy of external beam radiation therapy at doses of 8-15 Gy. It is commonly used for tracheobronchial obstruction but can potentially lead to life-threatening bleeding and airway erosion, requiring surgical intervention.

Chen et al reported that postoperative radiation therapy was only used for patients with positive margins and observed a significant improvement in overall survival and disease-free survival in these patients compared to those who received only incomplete resection without radiation therapy.

A study conducted by Bittner et al between 1989 and 2005, reported 20 patients with adenoid cystic carcinoma treated with fast neutron radiotherapy at the University of Washington. They considered it to be an effective treatment for locally advanced adenoid cystic carcinoma that may offer therapeutic benefits over commonly used treatment modalities. They reported a 5-year overall survival rate of 89.4% and a 5-year local control rate of 54.1%, primarily in patients with unresectable disease or locally advanced disease.²⁴ Neutron radiation therapy has proven to be effective in advanced or unresectable carcinoma and has been used in single-institution experiences with low morbidity rates, although some cases of tracheal cartilage stenosis or necrosis have been described.

In their case series, Levy et al described acute toxicity related to moderate-grade radiation therapy in all their patients. They reported esophagitis in 42% of cases, dysphonia in 32%, and mucositis in 9%. One patient experienced tracheoesophageal fistula during treatment. With regard to late toxicity signs, 7 patients (23%) developed symptomatic tracheal stenosis, and 5 (12%) required subsequent tracheotomy. Grade 3 dyspnea occurred in 4 patients (14%); and 5 patients (16%) developed hypothyroidism. 4 patients (12%) showed pericarditis. In the study of Chen et al, the most common adverse reactions were tracheitis and esophagitis.

Major complications following conventional radiotherapy, such as chondronecrosis, usually occur between 3 and 12 months after treatment.

Doggett et al performed percutaneous implantation of radioisotope seeds guided by computed

tomography in three patients diagnosed with adenoid cystic carcinoma. Two of them had previously undergone tracheal resection, laser ablation, and post-operative radiotherapy, and one patient declined resection and radiation therapy. All three patients responded well in the short term and during nine months of follow-up without chronic adverse effects and with reduced or relieved coughing. However, long-term follow-up is necessary to assess efficacy and toxicity.¹²

Chemotherapy

Few studies have shown the role of chemotherapy in the treatment of tracheal adenoid cystic carcinoma, and further studies are needed to clarify its efficacy. Chemotherapy does not have a significant role as primary therapy, but it may be considered for palliative treatment in cases of distant metastatic disease, for radiosensitization, or in combination with radiation therapy for unresectable carcinomas.

Cisplatin-based chemotherapy has been used successfully in a patient with an unresectable tumor in combination with radiation. However, this form of treatment has not been prospectively evaluated yet in primary tracheal tumors.

Tracheal adenoid cystic carcinomas are generally considered chemoresistant. Chemotherapy and targeted therapies administered alone are not indicated for localized tumors. Responses to chemotherapy regimens based on cisplatin, cyclophosphamide, and adriamycin have been evaluated.

Alternative treatments such as brachytherapy, photodynamic therapy, and cryotherapy are available but have not shown significant long-term benefits and are primarily used for palliation.

CASE REPORT

We present the case of a 70-year-old female patient with a history of arterial hypertension, former smoker (34 pack-years), diagnosed with COPD who presented with a hemoptysis episode. She had previously experienced dyspnea classified as functional class III-IV, which was managed as an outpatient exacerbation of COPD. A chest CT scan was requested, revealing a soft tissue density lesion located in the distal third of the trachea (Fig. 1 and 2), causing a narrowing of its lumen.

Tumor resection was performed through rigid bronchoscopy, revealing 80% obstruction of the tracheal lumen in the distal third area, associated with malacia and mucosal infiltration extending to both main bronchi. Two lesions were removed, one of 4×1.2 cm and another one of 1.4×1.2 cm (Fig. 3). The histopathological report of the lesions showed tracheal mucosa infiltrated by an atypical proliferation consisting of cribriform, tubular, and solid nests, lined by a biphasic population of inner cells with eosinophilic cytoplasm, round nuclei, and granular chromatin. There was an outer layer of cells with clear cytoplasm, round and oval nuclei with occasional isolated nucleoli, and basophilic



Figure 3. Surgical sample obtained with rigid bronchoscopy.

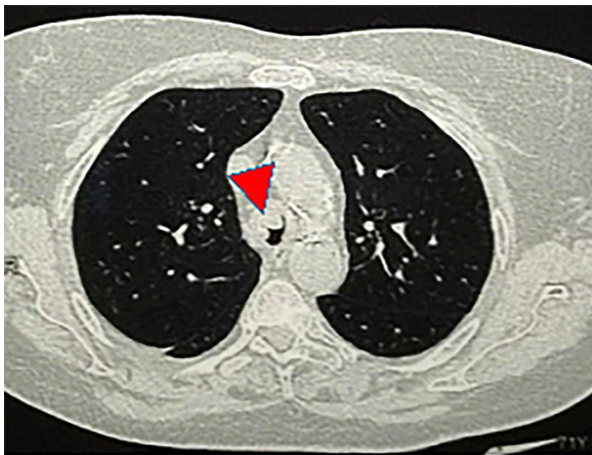


Figure 1. Lung window. Image with soft tissue density that causes a reduction in the tracheal lumen (arrowhead).

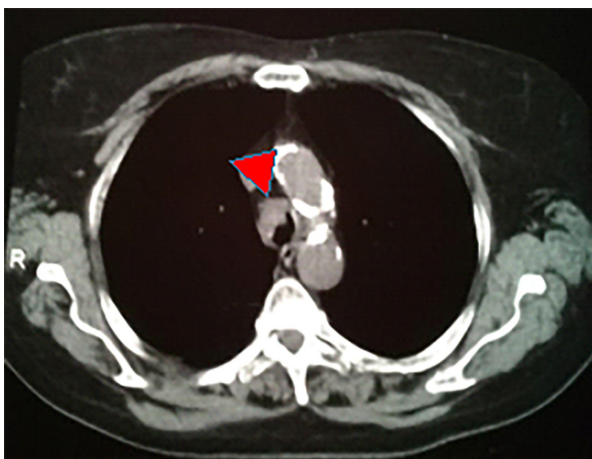


Figure 2. Mediastinal window. Image with soft tissue density in the distal third of the trachea (arrowhead).

intraluminal mucoid material. Positive AE1/AE3, negative TTF1 (thyroid transcription factor 1), S100 nuclear and cytoplasmic staining in myoepithelial cells, positive AML (acute myeloid leukemia) in myoepithelial cells, positive CALPONIN in myoepithelial cells, and positive CK7 in the inner layer, consistent with adenoid cystic carcinoma with resection margins compromised by the lesion.

The case was presented in a medical conference with the oncology department, where surgical resection was ruled out due to mucosal extension of the lesion to the carina and both main bronchi. A tracheobronchial Y-stent was placed, and the patient was clinically and endoscopically followed-up, on a regular basis.

DISCUSSION

Adenoid cystic carcinoma is a malignant tumor of the salivary glands that is relatively common in the head and neck region. However, its presence in the airway is rare.

The mean age of presentation is under 50 years, but the patient of this case was diagnosed at the age of 70, thus falling within the age range reported by Zhao et al, Calzada et al, Webb et al, and Chen et al. However, it correlates with the commonly described late diagnosis.

There is no gender predilection for the presentation of adenoid cystic carcinoma; however, Webb et

al, Chen et al, and Levy et al reported more cases in female patients.

No specific risk factors have been associated with the presentation of adenoid cystic carcinoma, as reported by Webb et al. The patient was a former smoker with a smoking cessation period of 35 years. Calzada et al found in their patient series that 36% of the cases were smokers.

The mean duration of symptoms before diagnosis is considered to be 18 months, with a range between 1 to 98 months. Dyspnea is the most common initial symptom, as reported by Zhao et al and Webb et al, so it can be underdiagnosed and confused with conditions such as asthma, COPD, or chronic bronchitis, thereby delaying the definitive diagnosis. The patient initially presented with functional class III-IV dyspnea, which, due to her clinical history, was initially considered as COPD exacerbation. Bronchoscopy was initially chosen as the therapeutic approach due to hemoptysis, since it is the second most frequent symptom following dyspnea, as reported by Webb et al.

Regarding the location, Webb et al, Chen et al, and Zhao et al described in their case series a predominance of tumors in the lower third of the trachea, similar to the patient described in this case. The macroscopic size of the tumors ranged from 1.5 cm to 8 cm, with an average of 3.1 cm, correlating with the macroscopic size of the tumor of our patient, which measured 4 x 1.2 cm; and had another one of 1.4 x 1.2 cm.

Tracheal adenoid cystic carcinoma is associated with very poor local and regional control, as demonstrated in 40% of patients in some series. Obtaining negative surgical margins is more challenging due to the relative inability to resect more than 6 cm of the trachea and the poor outcomes associated with tracheal grafts. Calzada et al reported that 80% of patients with adenoid cystic carcinoma had positive margins, predominantly in the distal trachea, and 40% had locoregional recurrences, which is consistent with the findings described by Zhao et al. The patient's histopathological report indicates that the resection margins are compromised by the lesion.

Calzada et al highlight in their study the tendency for local recurrences of adenoid cystic carcinoma in cases with positive margins and a distal location in the trachea, as is the case with the patient we

describe. During the follow-up period ranging from 4 to 168 months (average 31 months), two patients experienced local recurrence of the disease. One of them had a recurrence at 2 months, and the second at 1 month after surgery. One of these patients died at 16 months post-surgery, being the youngest in the series at 25 years of age.

Surgery is the cornerstone of treatment for adenoid cystic carcinoma; however, Ahn et al used laser removal through bronchoscopy in 2 out of 18 patients, which was successfully performed in selected cases of early-stage tumors. The patient came to the Emergency Service with hemoptysis. Endoscopic assessment and subsequent tumor removal through rigid bronchoscopy were performed.

In the review by Benissan-Messan et al, patients with adenoid cystic carcinoma were four times more likely to undergo resection, and survival was significantly higher for patients who underwent resection with curative intent. The overall mortality within 90 days following surgery was 2.5%, showing low perioperative mortality and a favorable long-term prognosis.

In their case series, Levy et al stated with regard to prognostic factors that the absence of perineural invasion and a dose of radiation therapy of ≥ 60 Gy correlate with better outcomes. Webb et al reported that 20 of 74 patients (27%) developed distant metastasis either as an initial presentation or during the follow-up period. Results were better for patients without lymph node metastasis or distant metastasis. Garden et al determined that the presence of perineural invasion of small nerves is not associated with worse control. Positive margins and involvement of major nerves were associated with an increased risk of local failure in patients treated with surgery and radiation.

Regarding mortality, Ahn et al did not find a significant difference between squamous cell carcinoma and adenoid cystic carcinoma but noted that pulmonary metastases were the primary cause of death in adenoid cystic carcinoma (6 out of 7 cases).

The 5-year mortality rate reported by Webb et al was 72.9%. Patients with adenoid cystic carcinoma and those with primary cervical tumors had better overall survival rates than other patients.

Zhao et al reported that survival after resection of all adenoid cystic carcinomas was 93.9% at 5 years and 61.1% at 10 years. In contrast, disease-

free survival was 73.9% at 5 years and 26.9% at 10 years.

Early diagnosis, experienced surgical treatments, and adjuvant postoperative radiation therapy for selected patients with positive margins can contribute to improving the survival of patients with primary tracheal adenoid cystic carcinoma.

The lack of a standardized staging system makes it difficult to compare studies, leading to a lack of advances in therapy or surveillance due to the rarity of these primary tumors. Therefore, multicenter studies are necessary to explore non-surgical future therapies that could be used as curative treatments for some patients.

Conflict of interest

Authors have no conflicts of interest to declare.

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