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Radiological Case Report / Caso Clínico

Primary Adenoid Cystic Carcinoma: An Uncommon Cause of Tracheal Obstruction

Carcinoma Adenóide Cístico: Uma Causa Rara de Obstrução da Traqueia

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Abstract

Adenoid cystic carcinoma (ACC) of the trachea is a rare malignant neoplasm, with little consensus in the literature regarding staging and therapeutic approach. Given the rarity of trachea primary neoplasms, it is sometimes difficult to make a correct diagnosis based on clinical and imaging findings, which often results in diagnosis delays. The radiologist's familiarization with this entity is essential for early detection and inclusion in the initial differential diagnosis of mediastinal lesions, guiding the remaining diagnostic and therapeutic approach.
We describe the case of a woman with clinical and imaging findings compatible with a tracheal neoplasm, that was confirmed to be an ACC. Given its longitudinal extension and the invasion of adjacent structures at diagnosis, it was considered surgically unresectable, and therefore palliative carbon-ion radiotherapy was proposed.

Keywords

Cystic adenoid carcinoma; Tracheal tumors; Tracheal stent; Staging; Radiotherapy.

Resumo

O carcinoma adenóide cístico (CAC) da traqueia é uma neoplasia maligna rara, pouco consensual na literatura em relação ao estadiamento e abordagem terapêutica. Dada a raridade das neoplasias traqueais primárias, é por vezes difícil o diagnóstico correto com os achados clínicos e imagiológicos, o que condiciona frequentemente atraso no diagnóstico. A familiarização do radiologista com esta entidade é fundamental para a sua deteção precoce e inclusão no diagnóstico diferencial inicial das lesões mediastínicas, assim como na orientação da restante abordagem diagnóstica e terapêutica.
Descreve-se o caso de uma mulher com achados clínicos e imagiológicos compatíveis com uma neoplasia primária da traqueia, que se confirmou ser um CAC da traqueia. Dada a sua extensão longitudinal e a invasão de estruturas adjacentes ao diagnóstico, foi considerada cirurgicamente irressecável, pelo que foi proposta para radioterapia paliativa com iões de carbono.

Palavras-chave

Carcinoma adenóide cístico; Tumores da traqueia; Stent traqueal; Estadiamento; Radioterapia.

Case Description

A 43-year-old female was referred for suspected lymphoproliferative disease. Two years earlier, in 2020, the patient had complaints of progressive dyspnea on minor efforts and coughing episodes, so a diagnosis of asthma was made in 2020 (respiratory function tests were compatible with severe obstruction). The patient did not smoke and had no other relevant medical history.
Due to recurrent episodes of hemoptysis and thoracic oppression, she went to the emergency department where a chest CT was performed, showing an intraluminal tracheal mass of soft-tissue attenuation, inseparable from the posterior wall of the trachea and centered at the level of the middle third of the trachea. The hypothesis of lymphoproliferative disease was first proposed.
A magnetic resonance imaging (MRI) was performed. It showed an oblong expansive retro-tracheal mass in the superior mediastinum (fig. 1), with slightly lobulated and well-defined contours, hypointense on T1-weighted images and hyperintense on T2- weighted images. On T1-weighted fat-saturated images, this lesion showed homogeneous enhancement after intravenous gadolinium administration. The mass showed invasion of the posterior wall of the trachea along almost its entire length with luminal obliteration greater than 50% and invasion of the esophageal wall at its most superior aspect. The MRI signal and morphologic characteristics of the lesion were suggestive of a tracheal neoplasm. Given the imaging findings and clinical data (43-year-old non-smoking female), the hypothesis of adenoid cystic carcinoma was proposed.
A positron emission tomography – computed tomography (PET-CT) was performed (fig. 2) which showed a high metabolic lesion posterior to the trachea. No other focus of high metabolic disease was found.
Ultrasound endoscopy with cytohistological sampling was performed (fig. 3). Immediately after the procedure, the patient had an episode of moderate hemoptysis and cardiorespiratory arrest, with reversal after advanced life support. In this context, she performed bronchoscopy for hemorrhagic control, which documented active hemorrhage in a wide-based vascularized mass (fig. 4). Given the risk of re-bleeding and the tracheal obstruction, it was decided to place a tracheobronchial stent (Novatech® GSSTM Y-STENT with 14/10/10 mm of outer diameters and lengths after adjustments of 9 cm for tracheal branch, 3 cm for left bronchial branch and 1.5 cm for right bronchial branch).

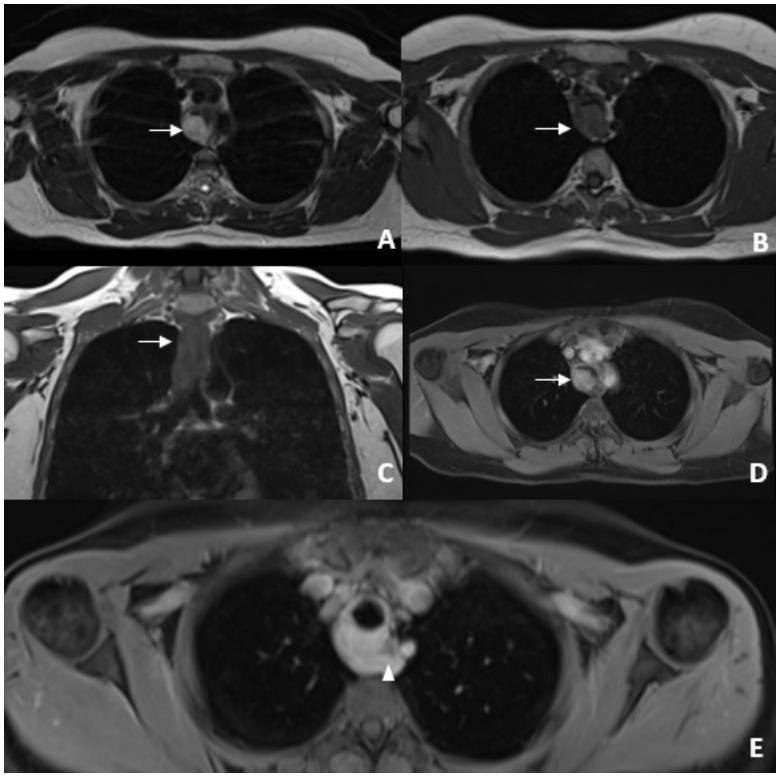


Figure 1 – Chest MRI axial T2-weighted image (A) shows a hyperintense oblong expansive retrotracheal mass with well delimited and slightly lobulated contours. Axial and coronal T1-weighted images (B and C) show a slightly hypointense lesion causing luminal obliteration greater than 50% (arrows). On T1-weighted images with fat saturation after contrast administration (D and E), this lesion demonstrated homogeneous contrast uptake. At the most superior aspect of the lesion, we see direct invasion of the esophagus (arrowhead).

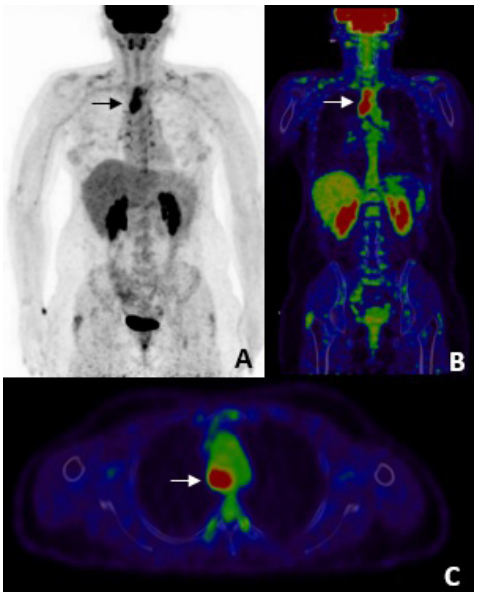


Figure 2 – 18F-FDG PET-CT with MIP (A) and fusion images in coronal and axial planes (B and C) showed hyperfixation of 18F-FDG (SUVmax = 10.77) in the mediastinal mass (arrows). No other relevant hypermetabolic changes were found.

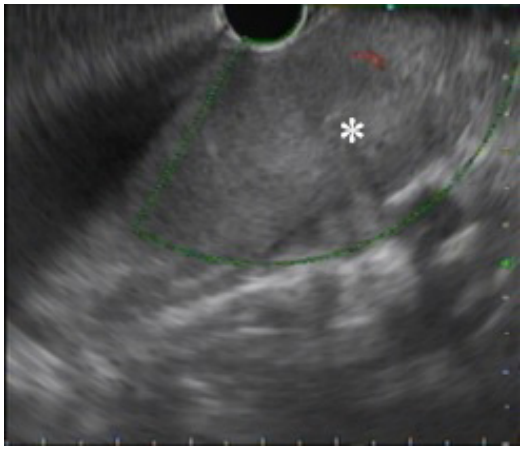


Figure 3 – Ultrasound endoscopic image demonstrating hypoechoic vascularized solid lesion (asterisks). Invasion of the esophageal wall was also noted.

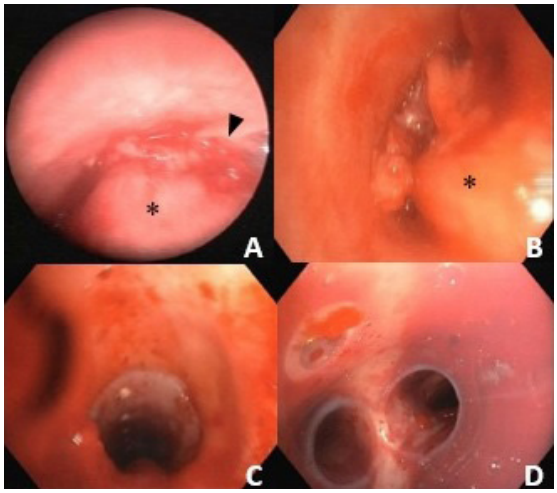


Figure 4 – Bronchoscopy shows a wide-based obstructing lesion in the posterior wall of the trachea (asterisks) with active bleeding (arrowhead) in images A and B. In the images C and D at the end of the procedure, a well-placed tracheobronchial silicone Y-STENT can be seen repermeabilizing the airway.

A chest computed tomography (CT) was performed due to chest pain and dyspnea maintenance, and it showed the tracheobronchial stent correctly placed, without associated complications (fig 5).

In the collected sample, bronchial mucosa was observed with extensive involvement by epithelial phenotype neoplasia, whose cytological and histological characteristics were compatible with salivary gland type neoplasia (ACC) (fig 6). The sample also showed a rearrangement involving the MYB gene, supporting the diagnosis.[1]

Although there is still no consensus on a staging system, according to some proposals described in the literature based on TNM tagging system, the patient was staged as cT4N0M0 given the signs of invasion of the adjacent esophageal wall.[2,3]

Considering the clinical staging and the longitudinal extension of about 8 cm (greater than 50% of the length of the trachea), the neoplasm was considered unresectable, so carbon-ion radiotherapy was proposed in Heidelberg, Germany.

In the follow-up, 8 months after radiotherapy, a marked reduction in tumor volume was observed. The tracheal stent was removed without complications and no suspicious lesions were identified at bronchoscopy. On serial CT and MRI imaging studies, the patient maintained stability of a slight upper and posterior peri-tracheal tissue thickening.

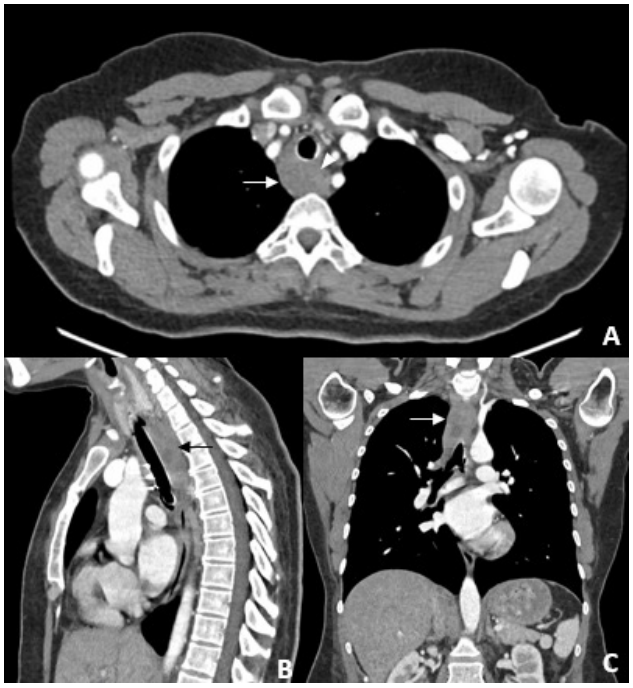


Figure 5 – Multiplanar images of chest CT (A, B and C) showed the tracheobronchial stent, with proximal end inferior to the cricoid cartilage and two distal ends in the main bronchi, with all segments permeable and no signs pneumomediastinum or pneumothorax. An infiltrative mass is observed circumferentially involving the trachea, with infiltration of the retro-tracheal mediastinal fat (arrows). The lesion does not show a cleavage plane with the right anterolateral wall of the proximal and middle thirds of the esophagus (arrowhead), which is compatible with suspected esophageal invasion documented in previous exams. No lymphadenopathy or lesions suspected of secondary involvement were identified.

more frequently slight or moderate uptake of iodinated contrast product, with polypoid or broad-based morphology with plaque-like or circumferential involvement and may have intra-luminal, intra-mural growth and/or extra-tracheal extension. Longitudinal extension is usually the longest axis of the lesion, being a characteristic finding of ACC and crucial to the therapeutic decision.[5,9] The tumor may have well-defined, lobulated or irregular margins and the existence of calcifications is rare. ACC can arise in the trachea, main bronchi, lobar bronchi, and more rarely in segmental bronchi. Perineural invasion is also characteristic.[9] About 10% of patients have lymphatic involvement or metastases at diagnosis, most of them in the lungs.[3] Magnetic resonance imaging can also help to delineate tumor extension.[10]

Bronchoscopy is frequently used to collect histological samples, thus allowing confirmation of the diagnosis. In our case, sample collection was performed through echo-endoscopy, which was additionally useful to corroborate previously suspected esophageal invasion.

There is no established staging system, but some groups have proposed systems based on the TNM staging.[2,3]

Segmental surgical resection with end-to-end anastomosis is the best therapeutic strategy in localized disease. Longitudinal extension greater than 50% of the length of the trachea precludes the surgical approach, markedly reducing the prognosis of our patient.[3,11]

The existence of positive margins after surgery is frequent, given the submucosal and perineural infiltrative behavior, justifying the frequent use of adjuvant radiotherapy.[5] Isolated radiotherapy is used in patients who are not surgical candidates.[11,12]

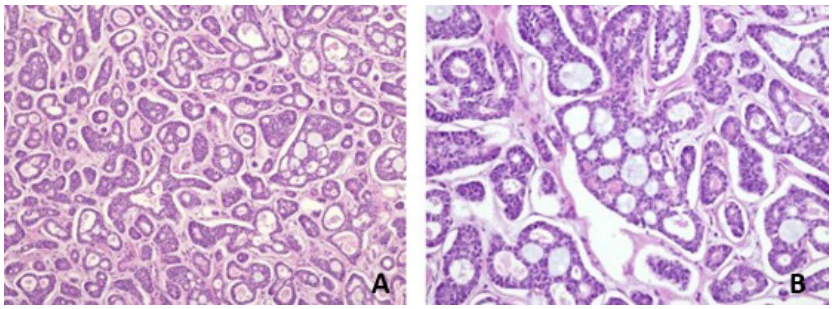


Figure 6 – Adenoid cystic carcinoma with cribriform and tubular pattern, H&E 100x (A) and x200 (B).

Discussion

Primary malignant tracheal tumors are rare.[4] In adults, the majority are squamous cell carcinomas (SCC), the second most frequent being ACC, approximately 30%.[5] Tracheal ACC is a low-grade, slow-growing malignant tumor that originates in the submucosal glands, occurring more frequently in the distal trachea.[6] Unlike SCC, ACC is associated with younger adults, with some female predominance, and there is no correlation with smoking.[4]

The most frequent clinical picture is related to airway obstruction, including cough, wheezing, dyspnea and, more rarely, hemoptysis. Given the nonspecific clinical presentation, patients are often initially misdiagnosed with other more frequent pathologies such as asthma,[7] as it happened in this clinical case.

CT is considered the best imaging approach, allowing localization, characterization and determination of its luminal and extra-luminal components, as well as the degree of tracheal obstruction, as demonstrated in the present clinical case.[8] Lesions are typically soft tissue masses with

Carbon ion radiation therapy is a recent and promising therapy option, being able to increase target doses while minimizing radiation exposure to adjacent organs at risk.[13]

This case highlights the difficulty at early diagnosis from clinic, respiratory function tests and imaging studies. The symptoms overlap with other diseases at an early stage such as asthma and the imaging characteristics can be easily mistaken with other most common lesions, thus delays in diagnosis and use of inadequate complementary diagnostic methods can occur. This context causes not only an increase in costs, but also a diagnostic delay that can make the lesion unresectable, with a very significant clinical impact on the prognosis of these patients.

In conclusion, radiologists and other medical professionals should be aware of tracheal neoplasms, including ACC. These lesions must be routinely and actively searched on imaging and should be considered in the differential diagnosis of mediastinal masses. ACC has some suggestive imaging features that should raise the diagnostic suspicion. Imaging is key for early detection, staging and resectability, which are critical to the therapeutic approach.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.
Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.
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Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.
Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.
Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.
Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).
Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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