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Tracheal Adenoid Cystic Carcinoma: A Case Report

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Abstract:
Adenoid cystic carcinomas (ACC), or cylindromas, are malignant tumors of the trachea that represent 0.1% of respiratory tract cancers. This case involves a 79-year-old female patient admitted for chronic dry cough, intermittent dysphonia and dysphagia, along with unspecified weight loss. The chest CT scan revealed circumferential thickening of the trachea with endoluminal budding. Bronchoscopy showed a vascularized tumor nodule at the distal part of the trachea, and the biopsy confirmed a tracheal adenoid cystic carcinoma. The PET scan showed a distal tracheal neoplastic process extending to the carina, with mild FDG avidity, and moderate subaortic and subcarinal mediastinal lymph node hypermetabolism. The patient received external radiation therapy. The outcome was marked by improvement in cough and dysphagia.

Keywords: Adenoid cystic carcinoma, trachea, radiotherapy.

Introduction:

Adenoid cystic carcinomas (ACC), or cylindromas, are malignant tumors of the trachea that represent 0.1% of respiratory tract cancers. They are the second most common primary malignant tumor of the trachea. We report the case of a patient admitted for chronic dry cough with circumferential thickening and endoluminal budding, revealing a tracheal adenoid cystic carcinoma.

Observation:

This is the case of a 79-year-old women with no significant medical history, who presented to the emergency department with a dry cough lasting for 2 years, intermittent dysphonia and dysphagia, without any other thoracic or extrathoracic signs, and evolving in the context of apyrexia and unspecified weight loss. Clinical examination revealed a performance status (PS) of 1, oxygen saturation of 96% on room air, with normal breathing and heart rate, and a normal pleuropulmonary exam.

The chest X-ray (Fig. 1) revealed widening of the intercostal spaces in the lower two-thirds of the right hemithorax, with a suspicion of narrowing of the tracheal lumen in its lower third. The chest CT scan (Fig. 2) showed circumferential thickening of the trachea with endoluminal mass. Flexible bronchoscopy (Fig. 3) revealed a large, vascularized tumor mass at the posterior part of the distal trachea, partially obstructing it, and topped by other smaller nodules. Biopsies were not performed due to the risk of bleeding and potential bronchial flooding. The carina appeared infiltrated and highly

vascularized, associated with a reduction in the caliber of both main bronchi, with the presence of nodules at the entrance of the right main bronchus.

Rigid bronchoscopy revealed the same endoscopic appearance, and the histopathological examination of the bronchial biopsies showed the presence of respiratory-type mucosa, occasionally malpighian in nature, hosting a carcinomatous proliferation. The first component consisted of basaloid cells with a myoepithelial appearance, with small basophilic cytoplasm and angular, hyperchromatic nuclei without nucleoli. The second component consisted of cuboidal epithelial cells with basophilic cytoplasm and oval, hyperchromatic nuclei, without nucleoli. The stroma was myxohyaline, with no perineural involvement or necrosis. The histological diagnosis was tracheal adenoid cystic carcinoma.

The PET scan showed a distal tracheal neoplastic process extending to the carina, causing narrowing of the lumen with mild FDG avidity, and moderate subaortic and subcarinal mediastinal hypermetabolism, suggestive of lymph node involvement.

The diagnosis was advanced, locally unresectable, non-metastatic tracheal adenoid cystic carcinoma.

Following the decision from the multidisciplinary consultation meeting, the patient underwent external radiotherapy with intensity-modulated radiation therapy (IMRT) at a dose of 66 Gy with conventional fractionation. The clinical outcome was characterized by cough and dysphagia improvement. The uniqueness of this case lies in the rarity of this histological type.

Discussion:

Tracheal adenoid cystic carcinoma is a rare primary malignant tumor, representing 0.1% of respiratory tract cancers, and is the second most common histological type of tracheal malignancies after squamous cell carcinoma [1]. These tumors are typically seen in younger individuals, with no gender predilection and no clear association with smoking [2]. Recent studies have shown that several genetic abnormalities predispose individuals to the development of these tumors. The EGFR pathway is involved in cell survival and proliferation. Overexpression of EGFR is frequently observed in up to 85% of cases, depending on the series [3, 4]. EGFR mutations are identified in 1-8% of ACCs [5, 6]. HER2 overexpression is found in 0 to 4% of cases [7, 8]. The proto-oncogene c-Kit is expressed in 80-90% of cases [9, 10].

The tumor most commonly develops from the secretory cells of the mucosa of the salivary glands, but can also affect the central airways (trachea, main bronchi), esophagus, nasopharynx, oral cavity, base of the skull, and very rarely presents as an endobronchial mass [11]. These tumors can be asymptomatic due to the large caliber of the trachea and the slow growth of the tumor, or they may present with clinical signs that are non-specific, such as dyspnea, stridor, wheezing, or an insidiously developing cough, which can lead to a misdiagnosis [12]. Chest X-ray may appear normal or show a laterotracheal opacity or an endotracheal tumor [12, 13].

Chest CT scan is essential for assessing the peritracheal extension and identifying potential secondary pulmonary or lymph node lesions [14]. Tracheobronchial fiberoptic examination is indispensable as it helps localize the tumor and confirm the diagnosis by performing biopsies, which are often difficult to interpret [12, 13, 14]. FDG-PET plays a diagnostic role in assessing lymph node involvement and detecting metastases.

Treatment for tracheal adenoid cystic carcinoma relies on surgery, radiotherapy, and interventional endoscopy. Surgery involves tumor resection with tracheal resection and termino-terminal anastomosis, followed by regional lymph node dissection. Lymph node dissection should not be too extensive to avoid compromising the tracheal vascularization. The approach varies depending on the tumor's location [15,

16].

Adjuvant radiotherapy is used to reduce local recurrence and can be used as first-line treatment to relieve symptoms if the tumor is non-resectable or if the patient is inoperable. Modern techniques, such as conformational radiotherapy and IMRT, help reduce the risk of high-dose exposure by minimizing the volume of tissue exposed. When external radiotherapy is used with a curative intent, a dose of at least 60 Gy is recommended, and if possible, up to 70 Gy in daily fractions of 1.8–2.0 Gy. A dose of 60 Gy is recommended for microscopic residual lesions in adjuvant treatment. Chemotherapy has no role except in metastatic cases [17, 18].

In cases of severe tumor obstruction, endoscopic laser resection allows for rapid deobstruction, followed by optimal local treatment [17].

Adenoid cystic carcinoma (ACC) is a tumor with attenuated malignancy, characterized by slow growth, progressive clinical evolution, a tendency for local recurrence, and late metastatic spread. Close and long-term endoscopic and radiological monitoring is necessary to detect any local or metastatic recurrence, which often involves the lungs, liver, lymph nodes, or bones. Its prognosis is better compared to that of squamous cell carcinoma, with a 5-year survival rate of 75% and a 10-year survival rate of 50%. According to Grillo et al., the median survival is 118 months after complete resection, 90 months after incomplete resection with adjuvant radiotherapy, and 28 months after exclusive radiotherapy [20].

Conclusion:

Tracheal adenoid cystic carcinoma is a rare tumor. Its diagnosis and surveillance rely on bronchoscopic examination combined with biopsy. The chest CT scan provides a better assessment of the extent and peritracheal spread of the tumor. Therapeutic management primarily involves surgery combined with radiotherapy.

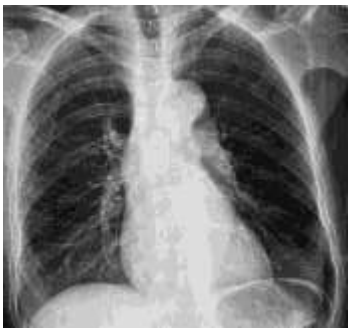
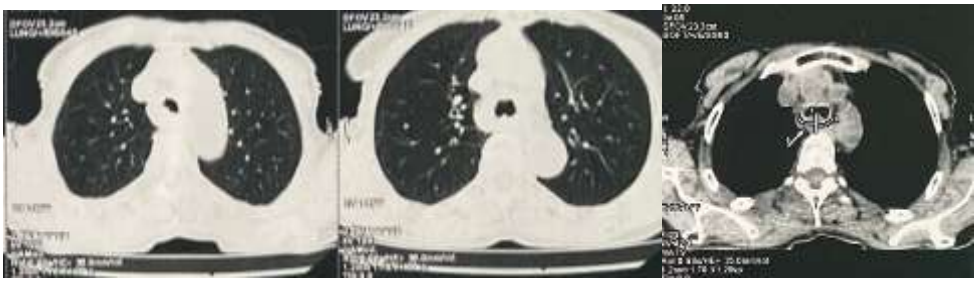


Fig. 1: Chest X-ray (frontal view): Widening of the intercostal spaces in the lower two-thirds of the right hemithorax, with suspicion of narrowing of the tracheal lumen at its lower third.



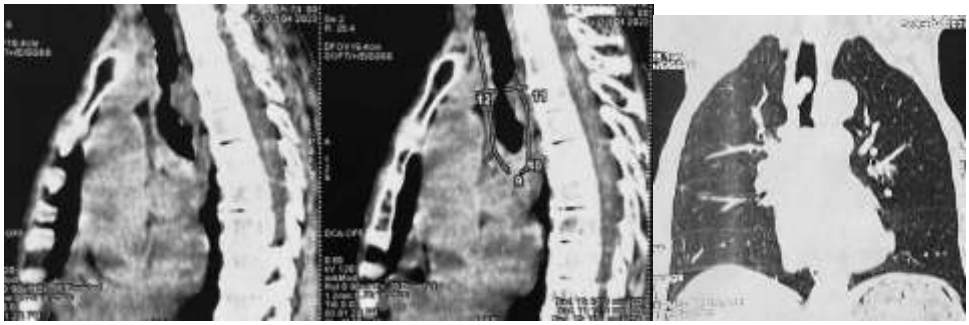


Fig. 2: Chest CT scan: Circumferential thickening of the trachea at its distal part, extending to the carina and segmental branches, with endoluminal budding (45 mm from the carina), narrowing its lumen. The maximum thickness measures 15 mm, extending over approximately 53 mm.

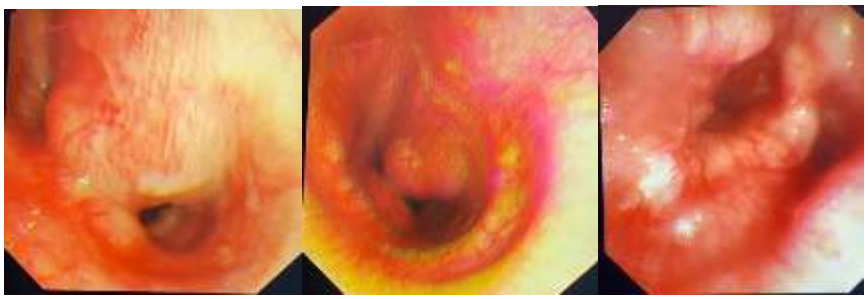


Fig. 3: Flexible bronchoscopy: A large tumor nodule at the distal part of the trachea on its posterior surface, partially obstructing the lumen, topped by other nodules. An infiltrated and highly vascularized carina with a reduction in the caliber of both main bronchi, and the presence of nodules at the entrance of the right main bronchus.

References

1. C. Le Péchoux, P. Baldeyrou, I. Ferreira, M. Mahé. Cylindromes thoraciques Cancer radiothérapie 2005, 9 : 358-361.
2. M. Soualhi, H. El ouazzani, J. Benamor, A. Chaibainou, M. El ftouh, S. Mouline, M.T. Fassy fihri, A. Benosman. Carcinome adénoïde kystique trachéal. A propos de trois cas. Rev Pno. clin., 2003, 59, 4 : 197-200.
3. Vered M, Braunstein E, Buchner A. Immunohistochemical study of epidermal growth factor receptor in adenoid cystic carcinoma of salivary gland origin. Head Neck 2002;24:632– 6. doi:10.1002/hed.10104.
4. Schneider T, Strehl A, Linz C, Brands R, Hartmann S, Beckford F, et al. Phosphorylated epidermal growth factor receptor expression and KRAS mutation status in salivary gland carcinomas. Clin Oral Investig 2016;20:541–51. doi:10.1007/s00784-015- 1541-1.
5. Saida K, Murase T, Ito M, Fujii K, Takino H, Masaki A, et al. Mutation analysis of the EGFR pathway genes, EGFR, RAS, PIK3CA, BRAF, and AKT1, in salivary gland adenoid cystic carcinoma. Oncotarget 2018;9. doi:10.18632/oncotarget.24818.
6. Dahse R, Driemel O, Schwarz S, Dahse J, Kromeyer-Hauschild K, Berndt A, et al. Epidermal growth factor receptor kinase domain mutations are rare in salivary gland carcinomas. Br J Cancer 2009;100:623–5. doi:10.1038/sj.bjc.6604875.
7. Glisson B, Colevas AD, Haddad R, Krane J, El-Naggar A, Kies M, et al. HER2 Expression in Salivary

Gland Carcinomas: Dependence on Histological Subtype. Clin Cancer Page | 13 Res 2004;10:944–6. doi:10.1158/1078-0432.CCR-03-0253.
8. Clauditz TS, Reiff M, Gravert L, Gnoss A, Tsourlakis M-C, Münscher A, et al. Human epidermal growth factor receptor 2 (HER2) in salivary gland carcinomas. Pathology (Phila) 2011;43:459–64. doi:10.1097/PAT.0b013e3283484a60.
9. Holst VA, Marshall CE, Moskaluk CA, Frierson HF. KIT protein expression and analysis of c-kit gene mutation in adenoid cystic carcinoma. Mod Pathol Off J U S Can Acad Pathol Inc 1999;12:956–60.
10. Jeng Y-M, Lin C-Y, Hsu H-C. Expression of the c-kit protein is associated with certain subtypes of salivary gland carcinoma. Cancer Lett 2000;154:107–11. doi:10.1016/S0304-3835(00)00387-6.
11. Charlton P, Pitkin L. Airway compromise due to adenoid cystic carcinoma obstructing the distal trachea: a review of current management and clinical trials. BMJ Case Rep 2015 [pii: bcr2014204063].
12. O. Bauduceau, B. Ceccaldi, O. Bernard, L.M. Dourhte, S. Le Moulec, R. Hervé. Le cylindrome de la trachée, tumeur rare d'évolution surprenante. Presse Med 2003; 32; n°13 : 602
13. A. Clought, P. Clarke. Adenoid cystic carcinoma of the trachea: A long terme problem. ANZ J.Surg.2006; 76:751-753.
14. Schneider P, Schirren J, Muley T, Vogt-Moykopf I. Primary tracheal tumors: experience with 14 resected patients. Eur J Cardiothorac Surg 2001; 20:12-8.
15. Le Péchoux C, Baldeyrou P, Ferreira I, Mahé M. Cylindromes thoraciques. Cancer Radiother. 2005 Nov;9(6-7):358–61. doi: 10.1016/j.canrad.2005.07.010.
16. Bauduceau O, Ceccaldi B, Bernard O, Dourhte LM, Le Moulec S, Hervé R. Le cylindrome de la trachée, tumeur rare d'évolution surprenante. Presse Med. 2003;32:602.
17. Muller A, Stokamp B, Schnabel T. Successful primary radiation therapy of adenoid cystic carcinoma of the lung. Oncology 2000; 58; 15-7.
18. Nakanishi, MD, PhD, Takemaru Kuruma, MD, Lizuka. Assisted thoracic tracheoplasty for adenoid cystic carcinoma of the mediastinal trachea. Surgery 2005; 137:250-2.
19. Jaso J, Malhotra R. Adenoid cystic carcinoma. Arch Pathol Lab Med 2011;135:511—5.
20. Grillo HC, Mathisen DJ. Primary tracheal tumors. Ann Thorac Surg. 1990;49:69–77. doi: 10.1016/0003-4975(90)90358-d.