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Primary Pulmonary Adenoid Cystic Carcinoma: A Case Report and Review of the Literature

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Abstract **Case Report**

Primary pulmonary adenoid cystic carcinoma (PACC) is a rare salivary gland-type malignancy arising from the submucosal glands of the tracheobronchial tree. It is generally characterized by slow growth and an indolent clinical course. Complete surgical resection with negative margins remains the preferred treatment for localized disease. However, management of locally advanced unresectable tumors remains challenging, and evidence regarding definitive chemoradiotherapy is limited. A 47-year-old man, with a 24-year history of snuff tobacco use, presented with a 6-month history of hemoptysis, dry cough, dyspnea. Chest CT revealed a 60 mm locally infiltrative left mediastino-hilar endobronchial mass causing ipsilateral pulmonary collapse, initially staged as cT3 N2b M0. Bronchoscopy showed a highly vascularized, completely obstructive tumor of the left main bronchus. Initial histological examination suggested a poorly differentiated non-small cell carcinoma. Immunohistochemistry demonstrated strong expression of CD117 and p63, with negativity for TTF-1, synaptophysin, and chromogranin A, supporting the diagnosis of adenoid cystic carcinoma. PET-CT confirmed locally advanced disease with bilateral mediastinal lymph node involvement and suspected carcinomatous lymphangitis. An otorhinolaryngological evaluation, including bilateral tonsillectomy, excluded a head and neck primary tumor. Surgical resection was considered unfeasible because of extensive local invasion with tumor encasement of more than 180° of the aortic arch. The patient therefore received four cycles of induction cisplatin and vinorelbine, resulting in significant tumor regression, with improvement of pulmonary atelectasis and mediastinal lymphadenopathy. He subsequently underwent definitive concurrent chemoradiotherapy, consisting of intensity-modulated radiotherapy to 60 Gy in 30 fractions with concurrent cisplatin and vinorelbine. This case highlights the diagnostic and therapeutic challenges of locally advanced unresectable PACC. Although surgery remains the cornerstone of treatment when complete resection is feasible, definitive chemoradiotherapy may represent a valuable alternative in unresectable disease.

Keywords: Primary pulmonary adenoid cystic carcinoma, Unresectable lung tumor, Definitive chemoradiotherapy, Induction chemotherapy, Immunohistochemistry, Case report.

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INTRODUCTION

Pulmonary adenoid cystic carcinoma (PACC), previously known as "columnar tumor", usually originates from submucosal glands of the trachea and bronchus. PACC is clinically rare and constitutes one of the major types of salivary gland carcinoma affecting the lung, although accounting for only 0.04%–0.2%. Unlike other bronchopulmonary tumors, adenoid cystic carcinoma (ACC) is not associated with smoking or other risk factors [1].

Primary adenoid cystic carcinoma of the lung is an unusual thoracic neoplasm with slow growing and low-grade malignancy. Usually, it is diagnosed at a higher clinical stage and is difficult to resect due to its central location. However, its clinicopathological features, demographics, treatment, prognosis, and long-term survival have not been fully elucidated due to its rare and unique nature [2].

In the management of primary pulmonary adenoid cystic carcinoma, radical surgical resection remains the cornerstone of treatment whenever feasible. However, for unresectable tumors, definitive chemoradiotherapy has emerged as a valuable therapeutic option, with encouraging local control and clinical outcomes reported in the literature.

CASE PRESENTATION

A 47-year-old male patient, with medical history of snuff tobacco use for 24 years, and no other family or personal history, was presented with hemoptysis, dry cough, dyspnea, weight loss, anorexia and fatigue, lasting for about 6 months. On clinical examination, the patient had a shortness of breath and stage II dyspnea. Chest auscultation revealed decreased breath sounds and decreased vocal fremitus over the left lung field, associated with crackles and wheezing. The superficial cervical lymph nodes were free.

A cerebral-thoracic-abdominal CT scan, revealed a locally infiltrative left mediastino-hilar endobronchial mass measuring 47x60x52mm (APxCCxTR), causing ipsilateral pulmonary collapse.

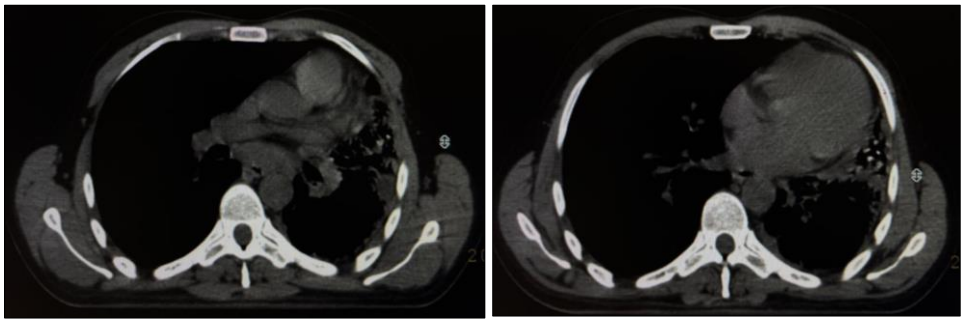


Figure 1: Transaxial CT scan showing a large left mediastino-hilar endobronchial mass

A bronchial fibroscopy was performed and evidenced a budding tumor in the left main bronchus, highly vascularized, completely obstructive. Multiple directed biopsies had been done for histological diagnosis.

Microscopically, the bronchial mucosa was infiltrated by a neoplastic proliferation composed of tumor cells with hyperchromatic and anisokaryotic nuclei and abundant eosinophilic cytoplasm. The tumor cells were arranged in a palisading pattern around perivascular spaces. Based on these findings, a poorly differentiated and infiltrative non-small cell carcinoma was initially suspected. An immunohistochemical study was therefore performed. The tumor cells exhibited a positive expression of CD 117 and P63 confirming the epithelial and myoepithelial differentiation. While no synaptophysin and chromogranin A, thyroid transcription factor 1 (TTF1) staining was identified. Taken together with the characteristic histological features, these findings supported the diagnosis of adenoid cystic carcinoma (ACC) of the left main bronchus.

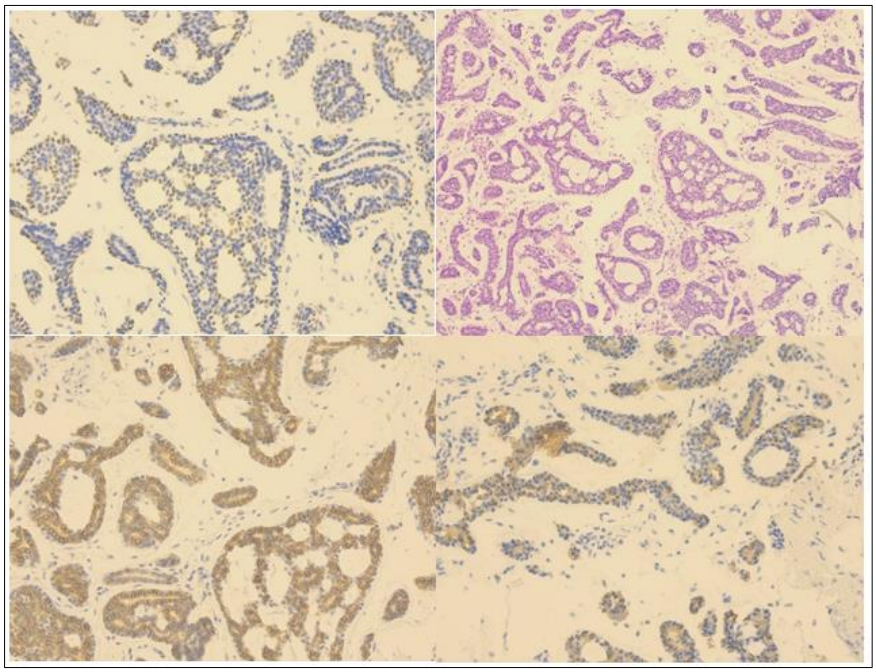


Figure 2: Histopathological and immunohistochemical features of adenoid cystic carcinoma. showing characteristic cribriform and tubular growth patterns with pseudocystic spaces containing hyaline material.

The PET-CT scan examination demonstrated a locally advanced left mediastino-hilar tumor with bilateral mediastinal lymph node involvement, associated with an appearance of carcinomatous lymphangitis of the left lung. In addition, fairly intense bilateral tonsillar hypermetabolism with bilateral hypermetabolic cervical lymphadenopathy were described. To identify a primary salivary gland tumor, a panendoscopy and a bilateral tonsillectomy were performed and were displayed no abnormalities. Based on the 9th edition of the TNM classification, the tumor was staged as cT3N2bM0. Surgical resection was deemed unfeasible because of extensive local invasion, with tumor encasement of more than 180° of the aortic arch.

Therefore, the patient received 4 cycles of induction chemotherapy with vinorelbine and cisplatin. The treatment regimen consisted of triweekly administration of vinorelbine (Navelbine 30 mg/m2) on days 1 and 8, while cisplatin 75 mg/m2 was administered on day 1 of each 21-day cycle.

A follow-up CT scan demonstrated a reduction in the size of the previously described left mediastino-hilar endobronchial mass, with regression of the ipsilateral pulmonary atelectasis and mediastinal lymphadenopathy, given an estimated response.

The patient subsequently underwent definitive concurrent chemoradiotherapy. Radiotherapy was delivered to a total dose of 60 Gy in 30 fractions of 2 Gy on the tumor and suspected hilar and mediastinal lymphnodes, concurrently with cisplatin and vinorelbine (same protocole).

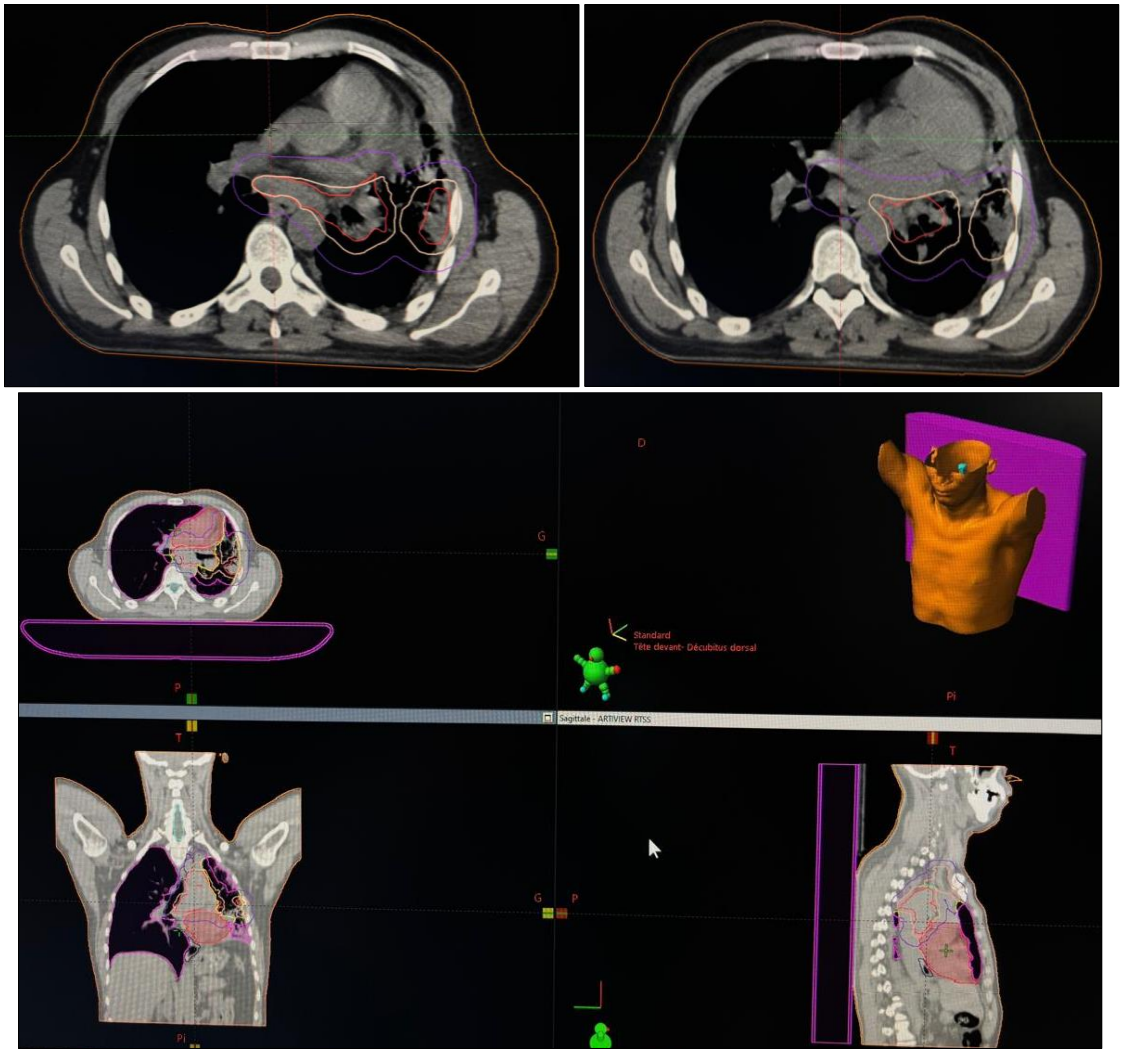


Figure 3: Radiotherapy treatment planning and dose distribution for locally advanced tracheobronchial adenoid cystic carcinoma

DISCUSSION

PACC is a rare but distinct salivary gland-type malignant neoplasm arising uncommonly as a primary tumor in the lung. It represents 0.04–0.2% of all lung cancers and can occur at any age, more commonly in the 40–60 age group, which was the case for our patient [1].

ACC of the lung has a slow growing, late metastasis and prolonged clinical course and hence, it is regarded as a low-grade malignancy. The site of origin is the trachea-bronchial glands in the airway submucosa, with a morphology similar to ACC arising in the salivary glands [2, 3].

Regarding the risk factors of PACC studies have reported that there is no significant relationship between PACC and smoking [4].

The clinical presentation of PACC patients largely depends on tumor location and distal obstruction. Cough with no obvious cause is the first symptom in most patients. Other symptoms, such as dyspnea, wheezing, hemoptysis, obstructive pneumonia were described. Therefore, it is easy to misdiagnose, leading to delay in treatment [2-5].

Histologically, PACC is classically characterized by three architectural patterns: cribriform, tubular, and solid. The cribriform pattern is the most characteristic [4].

In our patient, the initial biopsy raised the possibility of a poorly differentiated non-small cell carcinoma, highlighting the diagnostic difficulty associated with limited endobronchial specimens and the unusual presentation. In order to eliminate the diagnosis of conventional adenocarcinoma and small cell carcinoma of lung, the immunohistochemistry study is highly recommended. In fact, the expression of myoepithelial markers, such as P63, SMA, C-kit, and S-100 protein, is a strong argument for ACC of the lung. In our case, strong CD117, P63 expression and combined with negativity for TTF-1, synaptophysin and chromogranin A supported the diagnosis of PACC. Interestingly, Huang *et al*., reported a rare case of lung ACC exhibiting immunoexpression of CK, SMA, Calponin, C-kit, P63, and S-100 protein [6].

Cytogenetically, ACC is characterized by activation of the MYB pathway. The most obvious genomic characteristic of adenoid cystic carcinoma (ACC) is t(6;9)(q22-23;p23-24) translocation, resulting in the fusion of the gene encoding transcription factor MYB with the nuclear factor IB (NFIB) and leading to overexpressing MYB gene. However, the frequency of this alteration appears to be lower in pulmonary ACC than in head-and-neck ACC, with MYB-NFIB rearrangements reported in approximately 40% of pulmonary cases [4].

The mainstay of treatment for localized PACC is complete surgical resection with negative margins whenever technically feasible. Achieving an R0 resection is crucial because positive surgical margins are associated with poorer prognosis [7]. However, complete surgical resection may be impossible in centrally located, locally advanced tumors.

A clinicopathological study of 64 patients with PACC among which 53 patients underwent surgical resection, showed that 23 patients with R1 resection had a 5-year survival rate of 86%, and 31 patients with a R0 resection had a 5 year survival rate of 94% [8].

The role of radiotherapy in primary pulmonary adenoid cystic carcinoma (PACC) is better established in the postoperative setting, particularly in the presence of positive or close surgical margins, nodal involvement, or locally aggressive disease.

In unresectable tumors, definitive radiotherapy may represent a potentially curative or disease-controlling treatment option. Although the available evidence is mainly derived from retrospective series and case reports, PACC appears to be responsive to radiotherapy [9]. Several reports have suggested that the addition of concurrent platinum-based chemotherapy may provide a radiosensitizing effect and potentially improve local control.

Complete responses following definitive radiotherapy have been reported, including a case of tracheobronchial adenoid cystic carcinoma treated with 66 Gy in 2-Gy fractions, resulting in complete clinical and histological remission at 27 months. In another series, 10 patients who received concurrent chemoradiotherapy had a mean survival of 50.1 months [7].

In inoperable tumors, definitive radiotherapy has therefore been considered a treatment option, with high tumor response rates reported in some series. However, historically, survival outcomes have generally been less favorable than those observed in surgical series, with reported 5-year survival rates ranging from 12% to 27%. Interestingly, in the study by Lévy *et al*., patients treated with surgery did not demonstrate a significantly better prognosis than those treated with definitive radiotherapy, suggesting that definitive radiotherapy may provide meaningful long-term disease control in selected unresectable patients [7, 8].

More recently, definitive chemoradiotherapy has also been reported in unresectable tracheobronchial adenoid cystic carcinoma. A case report described a 30-year-old woman with unresectable tracheal adenoid cystic carcinoma treated with concurrent chemoradiotherapy using weekly paclitaxel and carboplatin, with a favorable tumor response. Two other limited reports have described combined chemoradiotherapy approaches with encouraging outcomes [9-12].

Primary radiotherapy data is less convincing, most likely because of selection bias, which has reserved radiotherapy for patients with the most advanced or unresectable disease or with significant medical comorbidities. Local regional control with primary

modern photon based radiotherapy or neutron beam radiotherapy is reported to be 56% and 57% at 5 years, respectively [13].

In a retrospective analysis, the results of chemoradiotherapy in 5 patients with adenoid cystic carcinoma treated with definitive chemoradiotherapy for reasons of unresectability or functional organ preservation. The patients were treated with a standard chemotherapy approach used for radiation sensitization. The results suggest that this approach offers an alternative to surgery followed by radiotherapy in selected patients [13].

The rationale for selecting induction chemotherapy in our patient was primarily based on the locally advanced and unresectable nature of the disease, characterized by extensive mediastino-hilar infiltration and major vascular involvement.

Although adenoid cystic carcinoma (ACC) is generally considered a relatively chemoresistant tumor, some cytotoxic regimens have demonstrated encouraging activity in advanced disease. In particular, the combination of cisplatin and vinorelbine has shown promising results in recurrent or metastatic salivary gland ACC, with an objective response rate of approximately 32% in the phase II study by Airoldi *et al*., [14]. Furthermore, this combination has been investigated in tracheal ACC, although tumor responses have been variable, highlighting the heterogeneous chemosensitivity of this histological subtype. In this context, induction chemotherapy was considered an individualized therapeutic approach aimed at achieving early disease control, assessing tumor chemosensitivity and potentially reducing the macroscopic tumor burden before definitive chemoradiotherapy [15]. Such tumor reduction could facilitate radiotherapy planning and potentially decrease high-dose treatment volumes, thereby limiting exposure of adjacent organs at risk [16]. However, the role of neoadjuvant chemotherapy in ACC remains poorly established, with no prospective evidence demonstrating a survival benefit, and it is not considered a standard treatment strategy [17].

Nevertheless, in selected patients with locally advanced unresectable disease, induction chemotherapy may represent a reasonable approach to optimize subsequent locoregional treatment.

However, these data remain limited, and no standardized chemotherapy regimen or optimal sequence of systemic therapy and radiotherapy has been established.

In our patient, surgery was considered unfeasible. He therefore received four cycles of induction chemotherapy followed with definitive concurrent chemoradiotherapy. The rationale for induction chemotherapy was to obtain early disease control and assess tumor response before proceeding to definitive local treatment, in orher to reduce the extend of radiotherapy volumes for less toxicity.

Although definitive chemoradiotherapy has been reported in unresectable tracheobronchial adenoid cystic carcinoma, the specific sequence used in our patient, consisting of induction cisplatin–vinorelbine followed by definitive concurrent chemoradiotherapy, appears to be poorly documented in the available literature. This case therefore illustrates a potentially feasible multidisciplinary approach for locally advanced unresectable PACC in the absence of a surgical option.

The patient has now completed his treatment and is currently under clinical and radiological surveillance.

CONCLUSION

Primary pulmonary adenoid cystic carcinoma is a rare malignancy for which diagnosis and management remain challenging, particularly in locally advanced unresectable stage. Although surgery remains the preferred treatment whenever feasible, definitive chemoradiotherapy represents a valuable alternative when complete resection cannot be achieved. Age, tumor size, lymph node metastasis status, margin status, and choice of treatment modality are independent prognostic factors that affect survival.

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