CASE REPORT

Primary Adenoid Cystic Carcinoma of the Lung: A Case Report of Rare Tumor

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Abstract

Adenoid cystic carcinoma (ACC) is predominantly a malignant neoplasm of the salivary glands but rarely can develop in the lungs as a primary tumor from bronchial glands, accounting for only 0.04%–0.2% of all pulmonary malignancies. Its slow growth, infiltrative nature, and nonspecific respiratory symptoms often mimic other conditions, creating considerable diagnostic and therapeutic challenges.

We report a case of a 50-year-old male radiotechnologist presented with recurrent hemoptysis and unintentional weight loss. Despite a normal chest X-ray, HRCT revealed a 4 × 3.1 cm soft tissue lesion completely obstructing the right principal bronchus with collapse of the right lower lobe. Pathological evaluation of bronchoscopic samples confirmed adenoid cystic carcinoma, necessitating a right lower bilobectomy. Subsequent histopathological examination revealed an intermediate-grade ACC with classic cribriform patterns, without lymphovascular and perineural invasion, and involvement of the middle lobe bronchial margin (stage pT1b pNx pMx).

Margin positivity raised concerns for potential recurrence, highlighting the need for meticulous follow-up and consideration of adjuvant therapy. This case emphasizes the importance of histopathological confirmation, margin assessment, and long-term surveillance in managing this rare pulmonary malignancy.

Keywords: Adenoid cystic carcinoma, Primary lung tumor, Endobronchial mass, Airway obstruction, Histopathology

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Introduction:

Adenoid cystic carcinoma (ACC) is predominantly a malignant neoplasm originating from the salivary glands but in rare cases, it may arise in the lungs-either as a primary tumor from the bronchial glands or as a metastasis from another site. This uncommon malignancy presents significant clinical challenges due to its atypical location, accounts for only about 0.04% to 0.2% of all lung malignancies.[1]

It arises from the submucosal seromucinous glands of the tracheobronchial tree, most often involving central airways.[2] Histologically, it resembles ACC of salivary glands, showing cribriform, tubular, and solid patterns with frequent perineural invasion. Notably, this carcinoma generally tends to grow along nerves (neurotropism), which leads to local spread and a high risk of recurrence. Although it can spread to other parts of the body, distant metastasis is rare.[3]

Patients frequently present with nonspecific symptoms such as cough, hemoptysis, or signs mimicking obstructive pneumonia, which can contribute to diagnostic delays.[4] The infiltrative nature of ACC renders surgical resection particularly challenging, and the treatment landscape remains largely undefined due to the rarity of the tumor and its unpredictable clinical course.

Here, we present a case of primary ACC of the lung in a middle-aged male, highlighting the clinicopathological features, diagnostic approach, and histopathological findings. The case is presented due to the tumor's unusual location, extreme rarity and potential for diagnostic delay.

Case report:

A 50-year-old male radio technologist from a low socioeconomic background presented with a 6-months

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history of recurrent hemoptysis and unintentional weight loss of approximately 2 kg. He had not experienced any shortness of breath, chest pain, cough, or fever. He was a nonsmoker but had a history of betel nut chewing and occasional alcohol intake. His family history was unremarkable. On examination, breath sounds were diminished over the right chest, with no supraclavicular lymphadenopathy; the remainder of the systemic examination was unremarkable.

The chest X-ray was normal, but the HRCT scan of the chest revealed a 4 × 3.1 cm soft-tissue lesion completely obliterating the lumen of the right principal bronchus and causing collapse of the right lower lobe. The remaining right lung parenchyma appeared normal. Bronchoscopy was performed, which revealed an endobronchial mass near the opening of the right upper lobe bronchus, causing complete luminal obstruction.

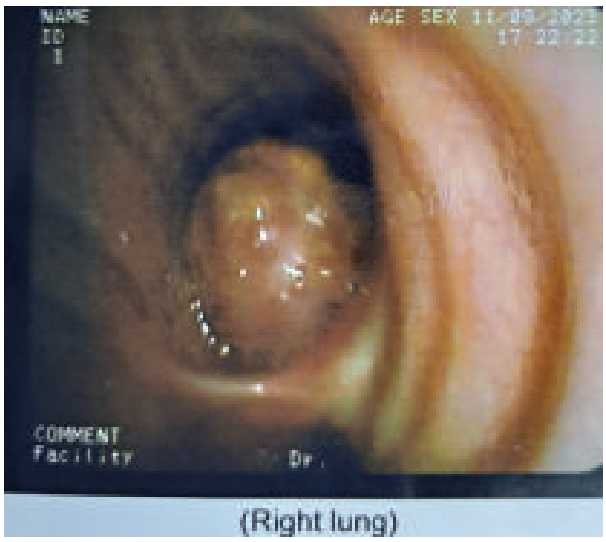

Fig: -1: *The bronchoscopy image shows a smooth, rounded endobronchial mass obstructing the lumen of the right bronchus.*

Pathological evaluation of bronchial washing (BAL), bronchial brushing, and bronchoscopic biopsy confirmed that the mass was malignant and favored adenoid cystic carcinoma. Subsequently, a right lower bilobectomy was performed.

The resected specimen included the middle and lower lobes of the right lung (14 × 7 × 5 cm). Gross examination of the middle lobe revealed an irregular solid gray-white mass measuring 2 × 1.8 cm, situated near the bronchial resection margin and 0.3 cm from the pleural surface. The rest of the middle and lower lobes showed dilated bronchi filled with fibrinopurulent exudate. Additional bronchial and endobronchial tissue appeared as multiple fragmented irregular gray-white pieces.

A histopathological examination with hematoxylin and eosin staining revealed intermediate-grade ACC. The tumor cells arranged in nests, cords, and cribriform patterns, with concentric arrangement around gland-like lumina containing homogeneous eosinophilic material. Lymphovascular and perineural invasion were absent. Areas of tumor necrosis were present, along with tumor-associated atelectasis and obstructive pneumonitis. The bronchial resection margin of the middle lobe was involved by tumor, whereas the visceral pleura and lower lobe bronchial margin were free of invasion. Additional lung parenchyma showed bronchiectasis but was uninvolved by tumor. Endobronchial tissue also demonstrated ACC with focal necrosis. The pathological staging was pT1bpNxpMx. All other investigations were normal. The patient was referred to oncology for further management.

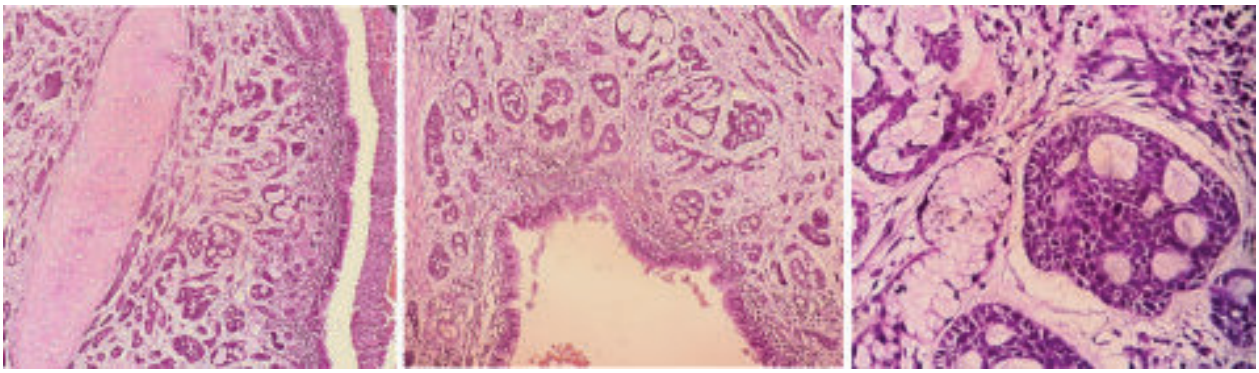

Fig:-2: Shows Intermediate-grade adenoid cystic carcinoma with cribriform growth patterns.

Discussion:

The patient's occupation as a radio technologist raises concerns about potential occupational exposures to ionizing radiation. Although he was a nonsmoker, his history of betel nut use and occasional alcohol intake may contribute to a multifactorial risk profile for lung tumors, although the specific etiological association with ACC is less well-defined in the literature.

The typical clinical presentation of ACC in the lung is often subtle, which can lead to delayed diagnosis. Patients typically report nonspecific symptoms such as cough, hemoptysis, or features of airway obstruction. In this case, the presentation was limited to recurrent hemoptysis without associated respiratory symptoms such as cough, dyspnea, or fever, making the initial diagnosis challenging.

Radiological imaging played a critical role in evaluation. While the chest X-ray was normal, HRCT revealed a soft-tissue density lesion obstructing the right principal bronchus, leading to further investigation. Bronchoscopy played a vital role in visualizing the endobronchial mass and obtaining tissue samples for pathological examination. The pathological examination of bronchial washing, brushing, and biopsy samples confirmed the diagnosis of adenoid cystic carcinoma, guiding the subsequent treatment plan.

Histologically, ACC is characterized by three architectural patterns—cribriform, tubular, and solid. The cribriform pattern is most common and generally associated with less aggressive behavior, whereas the solid subtype tends to be linked with higher-grade behavior and poorer outcomes.[4] In our case, the tumor exhibited intermediate-grade ACC featuring classic nests, cords, and cribriform arrangements, with eosinophilic luminal material, along with tumor necrosis and associated obstructive changes—aligning well with classic descriptions.

Surgical resection is the mainstay of treatment for localized pulmonary ACC, with reported 5-year survival rates of 85–91.7% in completely resected cases, compared to 0–33% in unresectable disease.[5] Margin status is a key prognostic factor, tumors with positive margins significantly increase recurrence risk. In this case, involvement of the bronchial margin highlights the need for close follow-up and consideration of adjuvant radiotherapy.

Pathological staging provides crucial prognostic and therapeutic information in pulmonary malignancies, including rare tumors such as adenoid cystic carcinoma (ACC). According to the 8th edition of the AJCC/UICC TNM classification for lung cancer, the tumor was staged as **pT1b pNx pMx,** indicating a tumor <2 cm confined to the lung without visceral pleural invasion.[6] This staging correlated with the findings in the resected middle lobe specimen.

The nodal status (pNx) could not be assessed in this case. Although lymph node involvement is less common in pulmonary adenoid cystic carcinoma (ACC) compared to other primary lung cancers, but its presence is associated with poorer outcomes. Thus, the absence of nodal sampling remains a limitation in staging accuracy.

The metastatic status (pMx) indicates that distant spread was indeterminate. Pulmonary ACC typically shows slow local progression with submucosal and perineural spread, while distant metastases to lung, brain, bone, or liver may occur at later stages.

Importantly, in ACC, conventional TNM staging does not always fully reflect prognosis. Margin status and growth pattern often have greater prognostic significance than tumor size alone.

In this case, despite the small tumor (T1b), the positive bronchial margin is a significant adverse feature, associated with higher risk of local recurrence. This highlights the need for long-term surveillance and consideration of adjuvant therapy in cases of incomplete resection.

From a pathogenetic standpoint, ACC is often driven by the MYB–NFIB gene fusion, a characteristic oncogenic event found in many cases. This genetic alteration is believed to drive tumorigenesis by upregulating MYB target genes involved in cell cycle progression, apoptosis inhibition, and angiogenesis.

Conclusion:

In summary, this case highlights the diagnostic challenge of primary pulmonary ACC due to its rarity, indolent presentation, and predilection for central airway location. Histopathological examination remains the gold standard for diagnosis, and complete surgical excision offers the best chance for long-term disease control. Given the potential for late relapse, extended follow-up is essential, even in early-stage disease.

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