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# Pulmonary Adenoid Cystic Carcinoma Presenting Late With Intrapericardial Extension: Case Report



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## ABSTRACT

Adenoid cystic carcinoma, also known as cylindroma, is one of the rare and unexplored clinical presentations of lung cancer, for which existing knowledge is scarce. This case report discusses a presentation of this tumor in the right lung, which subsequently extended to the left atrium through the right superior pulmonary vein. The extension of this rare tumor into the left atrium makes this case both uniquely distinctive and clinically relevant. The management strategy opted for this case was a right posterolateral thoracotomy and right pneumonectomy with partial resection of the left atrium. The desired outcome of this report is to shed light on the unusual clinical pathophysiology, register its atypical extensions, and navigate surgeons who may encounter this manifestation in the future.

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**Keywords:** Neoplasm; Atypical; Atrial resection; Case report

## Introduction

Lung cancers are known to have high mortality and morbidity rates globally.<sup>1</sup> They are broadly classified into small cell carcinoma (15% of the cases), which presents with neuroendocrine attributes and a high malignancy index, and NSCLC (85% of the cases), which have further pathologic subdivisions such as adenocarcinoma and squamous cell carcinoma.<sup>2</sup>

Adenoid cystic carcinoma (ACC), previously known as cylindroma, is a subtype of adenocarcinoma of the lung, affecting the lungs and associated upper airways. This rare salivary gland-type malignant neoplasm accounts for 0.04% to 0.2% of all primary lung tumors, making it a diagnostic and therapeutic challenge.<sup>3</sup> Extension into the left atrium is even rarer.

When this type of cancer invades the left atrium and right superior pulmonary vein, it is classified as locally advanced lung cancer.<sup>4,5</sup> To the best of the authors' knowledge, this is the first case to report the presentation of ACC extending into the left atrium from a research-deficient, low-middle-income country. Because of the rarity of its presentation, a holistic understanding of the clinical and pathophysiologic features, treatment

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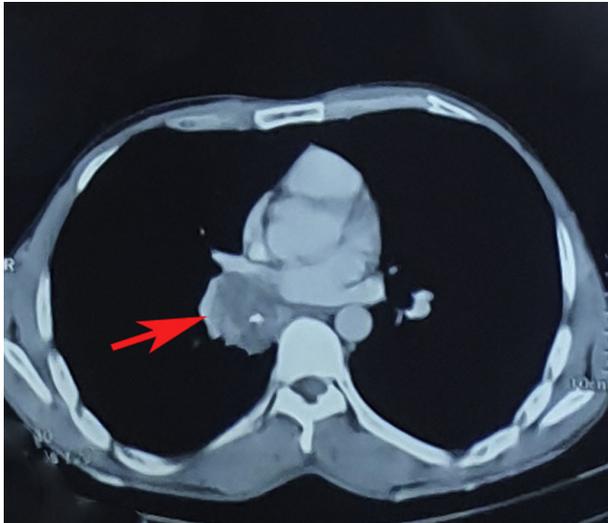
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**Figure 1.** Adenoid cyst visible on the right (mediastinal window).

modalities, and long-term prognosis are not very well established. However, few reports do state the 1-year and 3-year survival rates as 71.8% and 37.8%, respectively.<sup>4</sup>

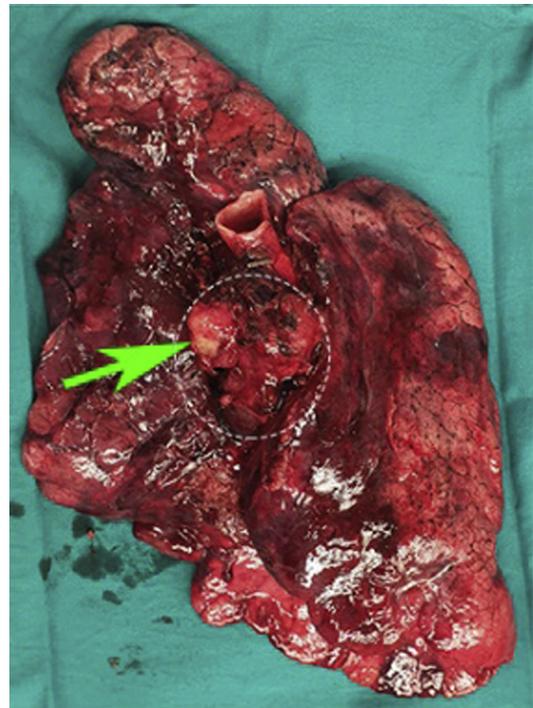
## Case Presentation

A 33-year-old man with no known comorbidities presented to the hospital with hemoptysis, preceded by chronic cough and intermittent chest pain for 4 years. Past medical history revealed that he had been diagnosed with pulmonary tuberculosis 7 years ago, for which he underwent successful treatment. Physical examination was unremarkable. Informed consent was taken from the patient to publish these findings in the literature.

An urgent computed tomography scan was done, which revealed a well-defined lobulated soft tissue density, with internal hypodensity indicating necrosis and coarse calcific specks involving the right hilar region extending up to the subcarinal location. It measured approximately 5.8 cm by 4.7 cm by 4.2 cm (craniocaudal × transverse × anteroposterior) dimension and involved the lung hilum at the level of the third thoracic vertebra (Fig. 1). There was considerable compression of the right mainstem bronchus and bronchus intermedius and loss of fat planes with the right pulmonary artery causing splaying of its branches.

The immunohistochemical examination of bronchioalveolar lavage revealed positive cytokeratin AE1/AE3 and focally positive p63. The endobronchial biopsy report confirmed the diagnosis of ACC of the right lung.

Surgical intervention was planned as the primary mode of treatment. The tumor site was approached



**Figure 2.** Posterior view of the left lung. Adenoid cyst in the hilar region.

through a right posterolateral thoracotomy and the right pleural cavity was entered. Exploration revealed that the tumor was densely adherent to the left atrium. On retrograde dissection, the following structures were sequentially divided: (1) inferior pulmonary vein, (2) the right mainstem bronchus, (3) the truncus anterior, and (4) the pulmonary artery. On entering the pericardium, the right and left atria and the pulmonary artery in that area were identified. The superior pulmonary vein was involved with the tumor, so the intrapericardial dissection of the left atrium was done. The left atrium was occluded with a partial occlusion clamp just distal to the entry of the superior pulmonary vein into the left atrium. The left atrium was cut and sewn in two layers with 4-0 Prolene. The tumor was removed with the entire right lung (Fig. 2). This was followed subsequently by lymph node dissection.

Histopathologic analysis of the excised tissue revealed lung tissue exhibiting a neoplastic lesion arranged in nodules and aggregates, and some areas revealed a solid pattern. The nodules and aggregates of the tumor revealed a predominantly cribriform pattern with cystic spaces containing basophilic material. These were lined by low cuboidal cells and surrounded by a myoepithelial layer. The cells contained eosinophilic cytoplasm with moderate to marked pleomorphic nuclei. Scattered mitotic activity was appreciated, and extensive areas of perineural invasion were identified. The tumor was 1.5 cm away from the bronchial resection margin and less than 0.1 cm away

from the outer painted pleural surface. A single lymph node was identified, which was tumor-free and was revealed to have anthracotic pigment.

Sections examined from the proximal vascular margin reveal fibrocollagenous tissue with vessels. There was no clear evidence of malignancy. Furthermore, the TNM staging described the tumor measuring 4.5 cm by 3.5 cm by 3 cm to be noninvasive (T2b) with no lymph node involvement (N = 0) and no metastasis (M = 0).

There were no intraoperative complications and the postoperative 1-year duration was unremarkable. No radiotherapy or chemotherapy was given after the surgery.

## Discussion

Primary ACC of the lung is a rare salivary gland-type malignant neoplasm that is distributed along the submucosa of the major airways.<sup>3</sup> ACC of lungs arises from the tracheobronchial glands distributed in the airway submucosa, with a histological appearance similar to ACC arising in the salivary glands.<sup>5</sup>

ACC occurs most often at other sites such as the breast, skin, uterine cervix, upper aerodigestive tract, and lung. The literature suggests that its extension to the left atrium is very rare. This type of tumor is seen in both men and women with a ratio of one-to-one, usually occurring in young adults, and is found more often in nonsmokers.<sup>3</sup>

Because it often presents with cough followed by hemoptysis and dyspnea and in patients who have a positive history of tuberculosis, ACC is often misdiagnosed as asthma or bronchitis.<sup>2</sup> Often, it may also remain asymptomatic until detected by imaging for other purposes such as routine evaluations.<sup>2</sup>

The literature suggests that lymphatic metastases are relatively uncommon, however, owing to the extensive spread along the major axis of the trachea at the time of diagnosis, residual tumors at the resection margin are not rare.<sup>3,4</sup>

The role of radiotherapy is not well defined in the literature and is only reserved for patients with either incomplete resection margins or those with unresectable disease.<sup>5</sup> This type of tumor also generally does not respond to chemotherapy but may exhibit a partial response to targeted novel therapies.<sup>3</sup> In some case reports, several chemotherapeutic agents including seven cycles of weekly paclitaxel combined with cisplatin, two cycles of docetaxel, and, subsequently, gefitinib were tried, but there was no response. Surgical resection seems to be the mainstay of treatment in these tumors.<sup>5</sup>

The strength of this report is the unique nature of clinical pathophysiology and the atypical extensions. The

scarcity of available literature is the limitation of the study, which makes it a valuable resource for surgeons around the globe.

## Conclusion

ACC presents a unique diagnostic challenge to clinicians and surgeons, owing to both a dearth of literature on it and the similarity of symptoms to other more known and understood lung pathologies. Once suspected, computed tomography scan followed by biopsy and histopathological analysis are vital next steps. Furthermore, once diagnosed, it is extremely important to resect all extensions of the primary tumor, as with the left atrium in this report.

## CRedit Authorship Contribution Statement

**Manzar Abbas:** Project administration, Writing - original draft, Writing - review & editing.

**Usama Qamar, Faiqa Binte Aamir:** Data curation, Writing - original draft.

**Syeda Maria Ahmad Zaidi:** Data curation, Writing - original draft, Writing - review & editing.

**Ali Aahil Noorali:** Writing - review & editing.

**Haseeb Ur Rahman, Saulat Hasnain Fatimi:** Conceptualization, Project administration.

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