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Original Research Paper

Pulmonary Medicine

A RARE CASE OF PULMONARY ADENOID CYSTIC CARCINOMA PRESENTING AS HYDROPNEUMOTHORAX

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ABSTRACT

Background: Pulmonary adenoid cystic carcinoma (PACC) is a rare malignancy, accounting for less than 0.2% of primary lung tumors. Due to its non-specific clinical presentation, it is often misdiagnosed as other respiratory conditions, leading to delayed diagnosis and treatment. The slow-growing nature of PACC and its tendency for airway obstruction further complicate early detection. Imaging and histopathology play crucial roles in confirming the diagnosis and differentiating it from other pulmonary malignancies. **Case Presentation:** We report a case of a 42-year-old male who presented with left-sided chest pain, dyspnea, and productive cough for one month. Initial imaging revealed left-sided hydropneumothorax, and further evaluation with high-resolution computed tomography (HRCT) demonstrated a soft tissue density mass with endobronchial extension into the left principal bronchus. Bronchoscopy identified a friable lesion obstructing the left bronchus, and histopathological examination confirmed adenoid cystic carcinoma via immunohistochemistry. The patient was managed with intercostal drainage, intravenous antibiotics, and planned for further oncological evaluation. **Conclusion:** This case highlights the importance of considering rare malignancies in persistent pulmonary symptoms, the role of imaging and histopathology in establishing the diagnosis, and the necessity of a multidisciplinary approach for optimal patient outcomes. Early diagnosis and intervention are crucial to improving prognosis in patients with PACC.

KEYWORDS :

INTRODUCTION:

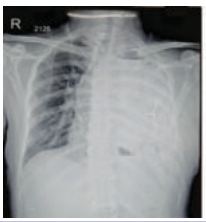
Pulmonary adenoid cystic carcinoma (PACC) is an extremely rare tumor arising from the tracheobronchial submucosal glands, accounting for only 0.04–0.2% of all primary lung cancers1. It is often slow-growing but exhibits a high propensity for perineural invasion and local recurrence2. Due to its rarity and overlap with common pulmonary conditions such as tuberculosis, pneumonia, and other lung malignancies, early diagnosis is often challenging3. Here, we present a case of PACC in a patient with prior pulmonary tuberculosis, initially misdiagnosed as a recurrent infection, emphasizing the importance of thorough clinical, radiological, and histopathological evaluation.

Case Presentation: A 42-year-old male presented to the emergency department at KLE Dr. Prabhakar Kore Hospital, Belagavi, with complaints of progressive left-sided chest pain radiating to the upper back, dyspnea worsening from mMRC grade 1 to grade 3, and a productive cough over one month. He also reported intermittent fever, generalized weakness, and loss of appetite. There was no significant weight loss, hemoptysis, or wheezing.

The patient had a past history of type 2 diabetes mellitus for six months and multiple episodes of tuberculosis (10 years back as Pulmonary , 2 years back as pleural effusion, and 6 months back). He had completed anti-tuberculosis therapy (ATT) from a government center in each instance. There was no history of COVID-19 or other significant comorbidities.

On clinical examination, the patient was hemodynamically stable, with oxygen saturation of 96% on room air. A left-sided intercostal chest drain (ICD) was noted in situ, draining turbid fluid with an active air leak. The trachea was shifted to the right, and fullness was noted on the left side. Percussion revealed hyperresonance in the left upper zones and dullness in the left basal regions. Auscultation demonstrated reduced breath sounds on the left side.

Investigations: A chest X-ray showed left-sided hydropneumothorax (Figure 1(a) &(b)). CECT thorax revealed a soft tissue density mass lesion with endobronchial extension into the left principal bronchus, resulting in partial obstruction with multiple specks of calcification in the mediastinal pleura and pleural thickening suggestive of neoplastic etiology (Figure 2, 3).



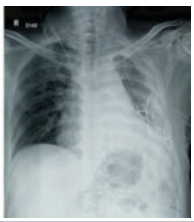


Figure 1 (a): CXR at presentation s/o Left sided gross hydropneumothorax
Figure 1 (b): Post ICD

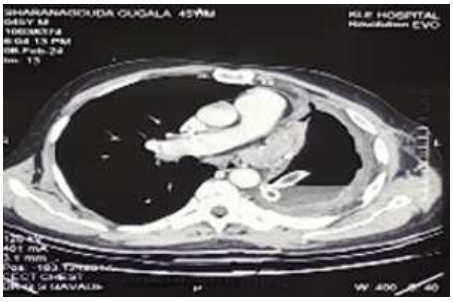


Figure 2

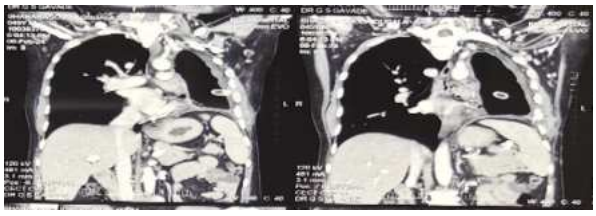


Figure 3

Figure 2 & 3: Heterogeneously enhancing soft tissue density mass lesion with multiple specks of calcification in the mediastinal pleura on left side with endobronchial extension into the left principal bronchus resulting in its obliteration with partial luminal narrowing of left pulmonary artery
Left ICD in situ

Bronchoscopy identified a friable, cauliflower-like lesion obstructing the left bronchus. Bronchoalveolar lavage (BAL) cytology was positive for malignant cells, and biopsy confirmed adenoid cystic carcinoma via immunohisto chemistry, positive for CK7, p63, EMA, and SMA (Figure 4).

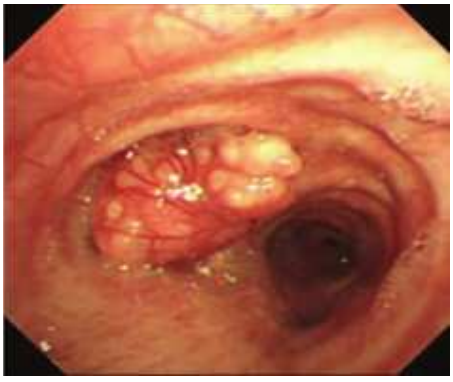


Figure 4: Bronchoscopic view showing a friable, cauliflower like lesion obscuring almost the entire left bronchus

Laboratory studies revealed an HbA1c of 11.2%, and neutrophil-predominant exudative pleural fluid with an ADA of 12 and LDH of 200 U/L. Sputum culture isolated Klebsiella pneumoniae sensitive to Piperacillin/Tazobactam, and BAL culture identified Staphylococcus aureus.

Treatment And Outcome: The patient was started on intravenous Piperacillin/ Tazobactam for empyema management and supportive care. PET-CT was planned for staging and further oncological intervention. However, due to financial constraints, the patient opted for palliative care and was subsequently lost to follow-up.

DISCUSSION: Pulmonary adenoid cystic carcinoma (PACC) primarily arises in the central airways, often leading to symptoms due to airway obstruction4. Its slow-growing nature and lack of strong association with smoking differentiate it from common primary lung cancers5. The histological hallmark includes cribriform patterns with mucinous or hyaline material. Immunohistochemistry plays a crucial role in distinguishing PACC from primary lung adenocarcinoma or squamous cell carcinoma6.

Management strategies include surgical resection for localized disease, with radiotherapy and chemotherapy as adjunctive therapies7. Given the high expression of MYB gene mutations in PACC, targeted therapies are emerging as promising treatment modalities8.

CONCLUSION:

This case underscores the importance of considering rare pulmonary malignancies in patients with persistent respiratory symptoms. Early diagnosis through imaging, histopathology, and immunohistochemistry is crucial for timely intervention and improved patient outcomes. A multidisciplinary approach involving pulmonologists, oncologists, and thoracic surgeons is essential for optimal management of PACC.

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