

Atypical endobronchial carcinoid tumor: A case report

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Abstract

Atypical Carcinoid Tumors are rare neoplasms derived from neuroendocrine cells. They are classified into two types: typical and atypical carcinoids. Atypical carcinoids are more aggressive than typical ones, with a higher risk of metastasis. They are distinguished by specific microscopic characteristics, such as a higher cellular proliferation index. The main treatment for endobronchial carcinoids is surgical, followed by close monitoring due to the risk of recurrence or metastasis.

We report the case of a 55-year-old man, with no significant medical history and no respiratory symptoms, who was diagnosed with an atypical endobronchial carcinoid tumor. The patient presented for preoperative evaluation related to gallstone disease, during which a chest X-ray revealed a left-sided thoracic opacity, leading to further investigations. A computed tomography (CT) scan showed an endobronchial mass, along with calcifications and mediastinal lymphadenopathy, which are characteristic features of atypical carcinoids.

Bronchoscopy was performed, revealing a tumor bud obstructing the left main bronchus. A bronchial biopsy confirmed the presence of atypical cells, supported by positive immunohistochemical markers such as chromogranin A and synaptophysin. The patient underwent a left pneumonectomy due to the obstruction and the tumor's aggressive potential, followed by adjuvant chemotherapy to reduce the risk of recurrence.

Postoperative follow-up was established with regular imaging studies to monitor for any potential recurrence. This case highlights the importance of early detection of atypical carcinoid tumors, even in asymptomatic patients, and the need for a multidisciplinary approach to ensure effective treatment and improved prognosis. Although rare, atypical carcinoids can be associated with a less favorable prognosis if not detected at an early stage.

Keywords: Atypical Carcinoid Tumor; Bronchoscopy; Pneumonectomy; Adjuvant Chemotherapy

1. Introduction

Bronchial carcinoid tumors, though rare, can be challenging to diagnose due to their often-asymptomatic presentation. Atypical carcinoids, in particular, have an increased potential for aggressiveness, requiring early identification for appropriate treatment. This case illustrates the importance of thorough evaluation in preoperative assessments.

2. Case Report

This is a case of Mr. R.E., a 55-year-old sports coach with no history of pulmonary tuberculosis and no recent exposure, and no toxic habits. He was under follow-up in visceral surgery for gallstone disease. The patient had no respiratory symptoms and presented to our facility following the incidental discovery of a left hilar-pulmonary tumor on chest X-

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ray during the preoperative work-up for his cholecystectomy. This finding was in the context of apyrexia and general condition decline.

On clinical examination, the patient was conscious, with a Glasgow Coma Scale (GCS) score of 15/15, WHO performance status of 0, respiratory rate of 18 breaths/min, oxygen saturation of 97% in ambient air, and heart rate of 87 bpm. Pleuro-pulmonary examination revealed a left-sided pleural effusion syndrome. The remainder of the examination was unremarkable. Chest X-ray (Figure 1) showed an opaque left hemithorax with mediastinal structure attraction.

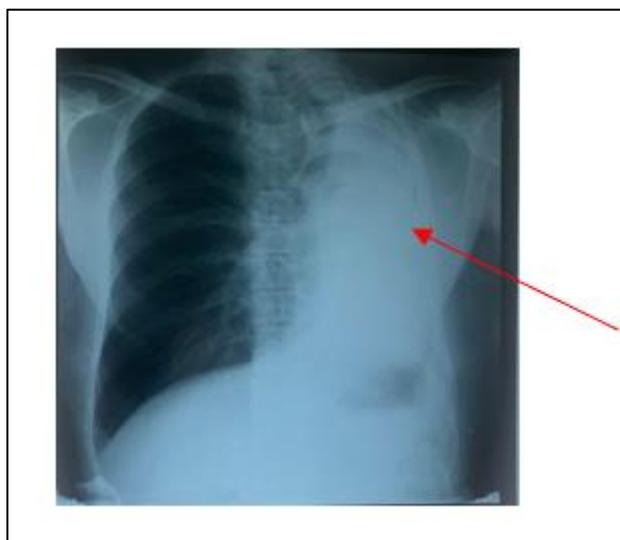


Figure 1 Dense, homogeneous opacity with a water-like density occupying nearly the entire left lung field, obscuring the diaphragm, the left heart border, and the corresponding recesses, with attraction of mediastinal structures towards the affected side. in relation to left lung atelectasis

A CT scan (Figure 2) reveals a left hilar-pulmonary lesion involving the left middle and dorsal basal segments, extending to the left main bronchus. The lesion is dense, heterogeneous, containing calcifications, and shows heterogeneous enhancement after contrast injection, measuring 82 x 76 x 123 mm. Pulmonary parenchyma shows areas of atelectasis with hypodense zones. Dense micronodules with well-defined borders are visible in the dorsal segment of the right upper lobe. Ipsilateral mediastinal lymphadenopathy and a small left pleural effusion are also present.

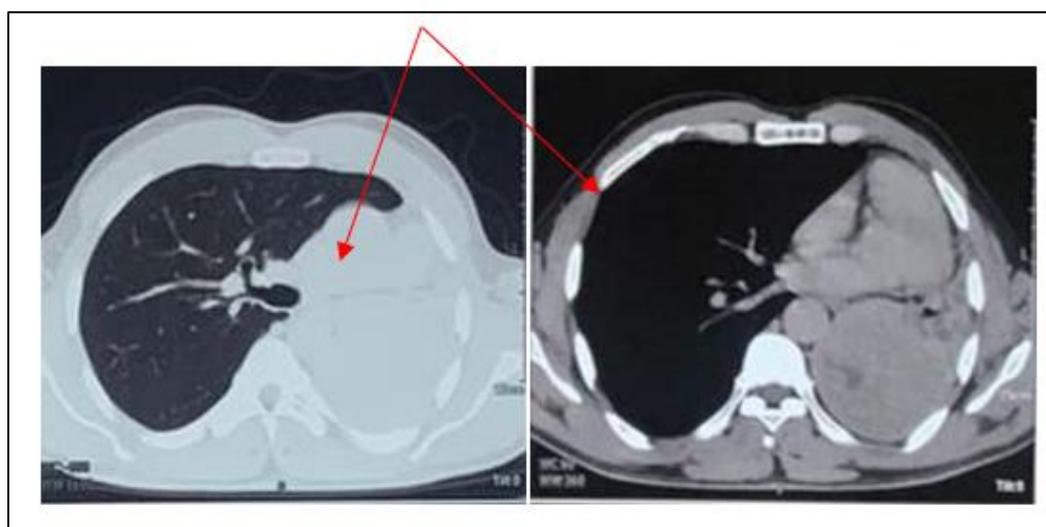


Figure 2 In the left parenchymal window: left hilar-pulmonary lesion involving the left middle and dorsal basal segments, extending to the left main bronchus. In the right mediastinal window: ipsilateral mediastinal lymphadenopathy and a small left pleural effusion

Bronchoscopy (Figure 3) revealed the presence of a pearly white tumor bud completely obstructing the left main bronchus, making it non-catheterizable, and bleeding easily upon contact.

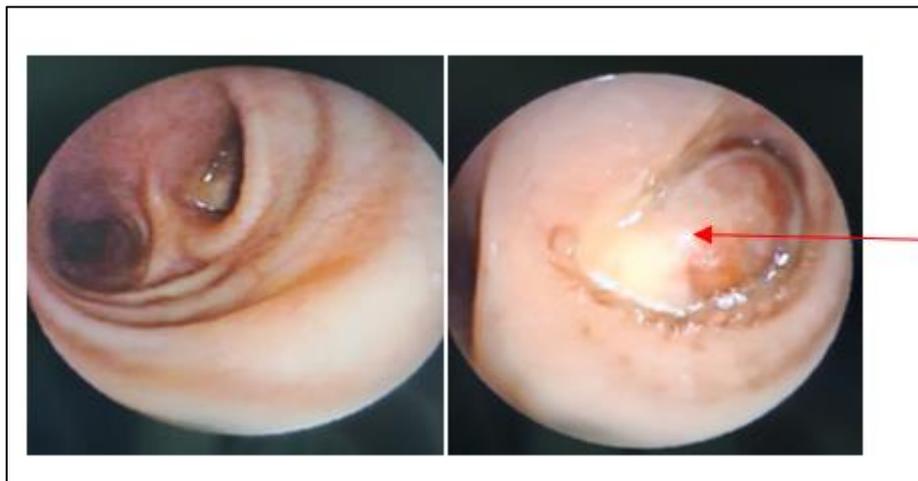


Figure 3 Pearly white tumor bud completely obstructing the left main bronchus

The biopsy of the tumor bud revealed: a morphological and immunobiological appearance consistent with a bronchial localization of an atypical carcinoid tumor (WHO 2020). The staging workup, including an abdominal-pelvic CT and a brain scan, was normal. Regarding the functional assessment: an ECG showed RRS, a heart rate of 65 bpm, a normal axis, and thin QRS complexes, with no repolarization abnormalities. Plethysmography revealed a moderate restrictive ventilatory defect with a Forced Expiratory Volume in 1 second (FEV1) at 67%. The 6-Minute Walk Test (6MWT) showed the patient was able to walk 450 meters, or 79.7% of the predicted distance, without desaturation.

The patient underwent a left pneumonectomy with adjuvant chemotherapy. The pathological examination of the surgical specimen revealed a morphological aspect of tumor proliferation with an endocrine-like architecture.

3. Discussion

Bronchial carcinoid tumors (CB) arise from neuroendocrine cells in the airways and are classified as neuroendocrine lung tumors, alongside typical carcinoids, atypical carcinoids, and large and small cell neuroendocrine carcinomas. These tumors are categorized based on their aggressiveness, from least to most aggressive (typical carcinoid < atypical carcinoid < large cell carcinoma < small cell carcinoma). Atypical carcinoids, accounting for approximately 10 to 20% of bronchial carcinoid tumors, have a higher malignant potential than their typical counterparts, making them more complex to manage clinically. They are characterized by a higher rate of cell proliferation (increased mitotic index) and may exhibit areas of tumor necrosis [1], [2].

Clinical symptoms at the time of presentation include nonspecific signs such as persistent cough, dyspnea, and recurrent pulmonary infections, with a high prevalence of neuroendocrine syndromes like Cushing syndrome and carcinoid syndrome. These syndromes can lead to the secretion of serotonin, histamine, bombesin, and other hormones, resulting in a wide range of hormonal symptoms. This clinical picture can complicate diagnosis, as these symptoms are not always immediately linked to a pulmonary tumor [3].

Chest imaging plays a critical role in detecting and characterizing bronchial carcinoids. In some cases, carcinoid tumors may be discovered incidentally during chest X-rays performed for other reasons. For example, a hemi thoracic opacity observed on a preoperative chest X-ray may prompt further evaluation via computed tomography (CT). CT scans allow visualization of the tumor mass, assessment of its extent, and examination for possible metastases or mediastinal lymphadenopathy. Tumoral calcifications are seen in about 30% of bronchial carcinoids and are an important radiological indicator [4]. An additional approach using magnetic resonance imaging (MRI) can be employed to more accurately evaluate soft tissues or lymphadenopathy. Furthermore, octreotide scintigraphy, which labels neuroendocrine cells with a somatostatin analogue, can detect regional or metastatic tumor spread [5].

The diagnosis of bronchial carcinoid is confirmed through bronchoscopy biopsy, where atypical cells are identified using a specific immunohistochemical profile. Markers such as chromogranin A and synaptophysin are used to confirm the

neuroendocrine nature of the tumor. However, unlike typical carcinoids, atypical carcinoids exhibit a higher mitotic index and greater cytological atypia, indicating a more aggressive tumor [6], [7].

Atypical carcinoids are characterized by a mitotic index ranging from 2 to 10 mitoses per 10 high-power fields, and tumor necrosis may be present, distinguishing them from typical carcinoids, which lack necrosis and have a low mitotic index. In terms of treatment, surgery remains the first-line option, aiming for complete tumor resection, with procedures such as lobectomy or pneumonectomy, depending on the tumor's location and extent. Once the tumor is removed, most neuroendocrine symptoms disappear [8], [9].

In more advanced cases or when aggressive features are present, adjuvant chemotherapy may be considered, although its efficacy remains debated. Some studies suggest it may reduce the risk of recurrence, especially in more advanced tumors [9]. Follow-up for patients after surgery is essential to detect any disease progression, particularly in the first five years postoperatively, and includes regular imaging studies and systematic clinical evaluations [10].

4. Conclusion

This clinical case underscores the importance of early detection of atypical carcinoids, particularly in asymptomatic patients. The use of systematic imaging, combined with a multidisciplinary approach, is crucial for improving prognosis. Regular and rigorous monitoring is necessary to quickly detect any recurrence and optimize long-term survival chances for patients with atypical carcinoids.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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