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(CASE REPORT)

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Unexpected tumor localization in a patient admitted for fibrotic ILD: The value of systematic bronchoscopic biopsy

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Abstract

Pulmonary lymphangitic carcinomatosis (PLC) is an aggressive form of pulmonary metastasis. Its fibrotic presentation can mimic interstitial lung disease (ILD), making diagnosis challenging when the primary tumor is unknown. We report the case of a 69-year-old patient admitted for the etiological assessment of chronic ILD at the fibrotic stage. While the initial clinical and radiological presentation suggested a classical interstitial pathology, systematic bronchial biopsies performed during bronchoscopy revealed poorly differentiated carcinomatous proliferation, with immunohistochemistry suggesting a pancreatico-biliary origin. This case highlights the crucial importance of systematic biopsy during bronchoscopy for any ILD, even when appearing fibrotic, to avoid overlooking an occult neoplastic pathology.

Keywords: Lymphangitic carcinomatosis; Bronchial biopsy; Pulmonary fibrosis; Adenocarcinoma; Differential diagnosis

1. Introduction

Pulmonary lymphangitic carcinomatosis (PLC) is an aggressive form of pulmonary metastasis resulting from the infiltration and obstruction of pulmonary lymphatic vessels by tumor cells [1]. It accounts for approximately 6% to 8% of all intrathoracic metastases [1, 3]. Due to its non-specific radiological semiology, PLC is frequently confused with other interstitial conditions such as idiopathic pulmonary fibrosis, sarcoidosis, or pulmonary edema [3, 6].

The clinical interest of this case lies in demonstrating how a terminal fibrotic lung presentation can mask an underlying occult neoplasia. It highlights the crucial importance of a systematic invasive diagnostic approach, including bronchoscopy with biopsies, to avoid diagnostic errors in patients presenting with fibrotic-appearing interstitial lung disease (ILD) [2, 5]. This approach is essential even in the absence of obvious macroscopic endobronchial lesions.

2. Case Report

A 69-year-old male patient, a merchant and former smoker, was admitted for the etiological assessment of a chronic ILD at the fibrotic stage. He presented with a 4-month history of progressive exertional dyspnea (Sadoul stage II), associated with an intermittent dry cough and a decline in general health marked by anorexia and weight loss.

Clinical examination upon admission revealed a conscious patient, eupneic at rest (17 breaths/min) with SaO2 at 98% on room air. Pleuropulmonary auscultation revealed "velcro" crackles at both bases and diffuse wheezing. There were no signs of right heart failure or digital clubbing.

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An AP chest X-ray showed diffuse bilateral reticulomicronodular opacities (Figure 1). High-resolution computed tomography (HRCT) demonstrated advanced pulmonary fibrosis with honeycombing, traction bronchiectasis, and irregular thickening of the interlobular septa (Figure 2). Initial laboratory tests revealed significantly elevated tumor markers:

Table 1 Results of Serum Tumor Markers

Tumor Marker	Patient Value	Reference Range
CA 19-9	> 12,000 U/mL	< 37 U/mL
CA 125	287.3 U/mL	< 35 U/mL
ACE	Normal	8 – 52 U/L

while angiotensin-converting enzyme and immunological tests were normal.

A flexible bronchoscopy was performed, showing inflammatory mucosa in both bronchial trees with thickening of the divisional spurs (Figure 3). Systematic bronchial biopsies of these spurs were performed. Pathological examination concluded an infiltration by a poorly differentiated non-small cell carcinoma. Immunohistochemical (IHC) study revealed a CK7+++ and focal CK20 positive phenotype, with negativity for TTF1 and P40, strongly suggesting an adenocarcinoma of pancreatico-biliary or upper digestive origin. A follow-up abdomino-pelvic CT scan did not show any obvious suspicious lesion, highlighting the occult nature of the primary tumor despite the massive pulmonary involvement.

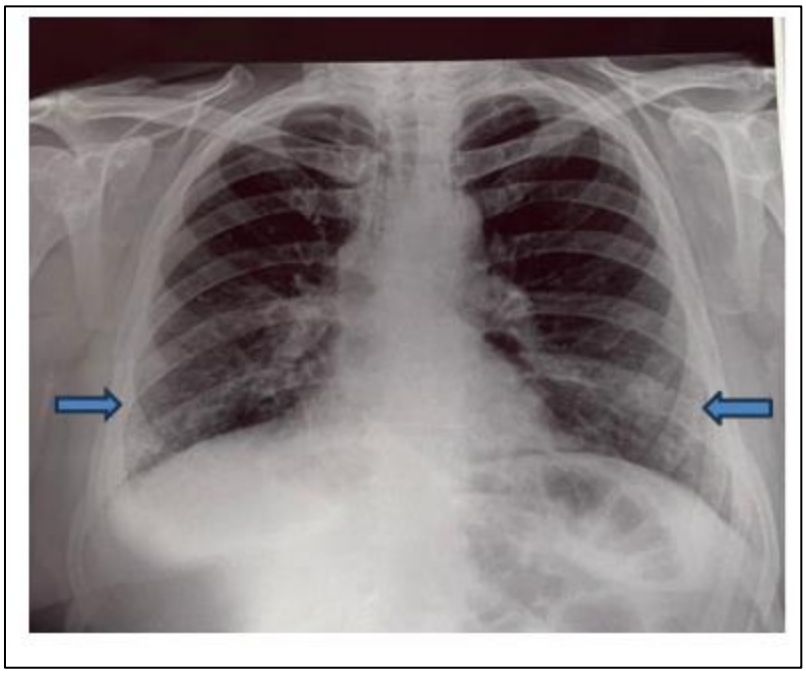

Figure 1 AP Chest X-ray showing diffuse bilateral reticulomicronodular opacities

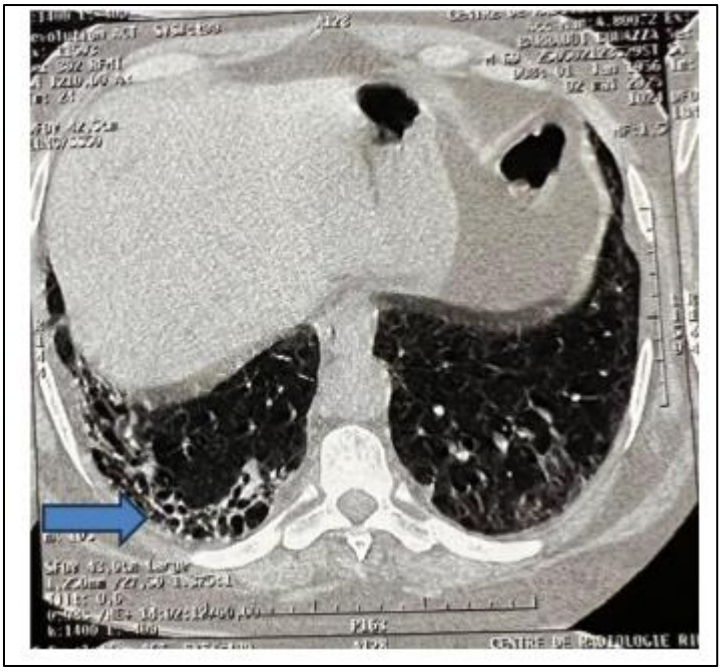

Figure 2 High-resolution chest CT (HRCT) scan demonstrating advanced fibrotic patterns with characteristic honeycombing and traction bronchiectasis

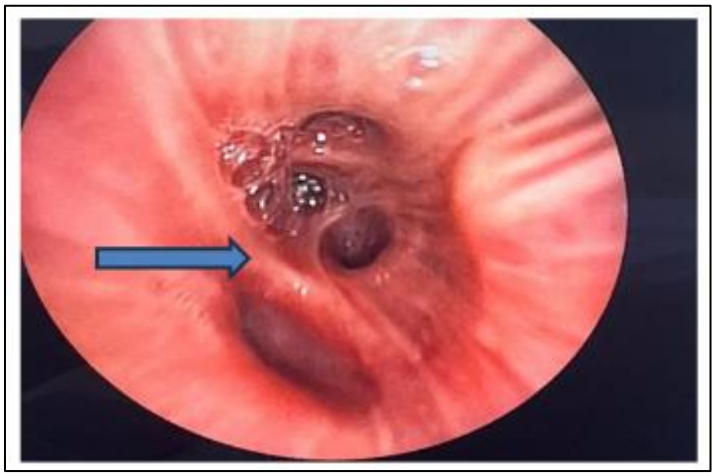

Figure 3 Endoscopic appearance demonstrating diffuse thickening of the bronchial spurs

3. Discussion

The diagnosis of PLC remains a major clinical challenge because its symptoms—progressive dyspnea and dry cough—are identical to those of chronic ILD [2, 6]. Radiologically, although nodular thickening of the septa and peribronchovascular sheaths is suggestive, at the fibrotic stage, these signs often merge with the architectural distortions of usual interstitial pneumonia (UIP) [3, 5]. As illustrated in this case, CA 19-9 can be a valuable marker, although its sensitivity remains limited in the absence of a visible mass on standard imaging [1, 3].

The use of immunohistochemistry is essential to differentiate a primary pulmonary tumor from metastatic disease, especially when the primary site is occult [4]. In our case, the CK7+/CK20+/TTF1- phenotype strongly suggests a glandular metastatic origin, such as the upper digestive or pancreatico-biliary tract, rather than a primary lung adenocarcinoma [4, 6]. This case confirms that systematic biopsy of the bronchial spurs maintains a high diagnostic value in PLC, as it can detect sub-epithelial lymphatic infiltration even when macroscopic endobronchial masses are absent [2].

Management of PLC at the fibrotic stage relies primarily on systemic chemotherapy tailored to the histological type [2]. The objective is to stabilize respiratory function and improve quality of life. As observed in our patient, a partial response can lead to the regression of pulmonary infiltrates.

However, the appearance of new vertebral bone lesions illustrates a "dissociated response," where one metastatic site improves while another progresses [6]. This necessitates a multidisciplinary approach, combining oncological protocols with supportive care, such as bisphosphonates or analgesics, to manage bone complications and preserve functional status [5, 6]. Once the fibrotic stage is reached, the overall prognosis remains grim, often requiring the early integration of palliative care [2, 6].

4. Conclusion

This case report serves as a reminder of the need to maintain a high clinical suspicion of malignancy in any ILD appearing fibrotic, particularly in the presence of a decline in general health. It emphasizes the importance of systematic investigation by bronchoscopy with biopsies, even in the absence of macroscopic endobronchial lesions, to avoid diagnostic errors and guide patients early toward oncological or palliative care.

Compliance with ethical standards

Disclosure of conflict of interest

The author declares no conflict of interest

Statement of informed consent

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

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