Progressive dyspnea due to pulmonary carcinoid tumorlets

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1. Introduction

Pulmonary Carcinoid tumors (PCTs) are nests of hyperplastic, neuroendocrine cells that extend beyond the basement membrane [1]. The diameter of nests differentiates tumorlets (<5 mm) from carcinoids (≥5 mm) [2]. They manifest as nodules in chest computer tomography (CT) [3]. Often coexist with other lung diseases, like fibrosis and bronchiectasis [4]. Literature suggests hypoxia, fibrosing process or genetic factors as underlying pathophysiological mechanisms.

1.1. Case history

A 77 years-old female presented in our Division of Pulmonology in the Department of Clinical Therapeutics of the National and Kapodistrian University of Athens School of Medicine, in Alexandra Hospital of Athens, Greece, with progressive dyspnea during the last month and non-productive cough. The woman suffered from hypertension, dementia, and gastric ulcer, under treatment with olmesartan, metoprolol, levodopa, rivastigmine, and omeprazole, respectively. She had not known previous pulmonary medical history. The auscultation revealed expiratory wheezing, with no other pathological signs. Arterial blood gases on room air proved a mild hypoxemia (pH 7.43, PaO2 72 mmHg, PaCO2 36 mmHg, HCO3- 22 mmol/l). Her spirometry was normal. A chest x-ray showed reticular opacities with small nodules. High resolution chest CT revealed alveolitis and “ground glass” opacities to the right lower lobe, fibrotic lesions and numerous micronodules, A thoracoscopy was performed and the obtained biopsy showed carcinoid tumors, with positive CK8/18, CD56, TTF-1 and synaptophysin immunohistochemical markers. Pulmonary carcinoid tumorlets are rare, benign tumors and individuals with tumorlets are typically asymptomatic. Our report presents a symptomatic clinical case of carcinoid tumorlet.
2. Discussion

Pulmonary Carcinoid tumorlets (PCTs) are rare, benign and usually constitute incidental findings [1]. They arise from focal proliferation of bronchial and bronchiolar neuroendocrine Kulitschitsky cells, which exceed basement membrane. They often coexist with diffuse bronchiectasis, emphysema and interstitial fibrosis.

Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH), tumorlets, and carcinoids are distinct entities that share a common initial event, the neuroendocrine cell proliferation [3]. PCTs and carcinoids have identical histologic findings and the diameter of the lesion is currently the only criterion discriminating them: <5 mm and ≥5 mm, respectively. Some carcinoids develop in patients with DIPNECH and multiple carcinoid tumors [5]. According to different studies the hyperplasia of pulmonary neuroendocrine cells can be an adaptive response to hypoxia, or a secondary process associated with pulmonary fibrosis, or a genetic predisposition [1,3,5].

Women, between 60 and 70 years old, are affected more than men (male to female ratio 1:4) [3]. Patients with tumorlets are typically asymptomatic. In a previous case report a carcinoid tumorlet in pulmonary sequestration with bronchiectasis was diagnosed during regular reexamination with x-ray, after breast cancer [6,7]. In another case report both tumorlet and adenocarcinoma were detected in the right upper lobe of a patient without symptoms or underlying lung disease [8]. Our patient claimed dyspnea and cough during the last month.

In a series of 294 patients with multiple carcinoid tumors clinical symptoms and significant airflow limitation were rare. Long-term survival was excellent, although patients had persistent disease [9]. No suggested therapies are described.

We describe this case report in order to highlight that tumorlets should be included in the differential diagnosis when a patient complain of dyspnea and small nodules are identified in the chest computer tomography. We should keep in mind that PET-CT is not the best method to diagnose this entity, however; targeted therapy with octreotide or 18F-DOPA amino acid analog can be used [10].

Conflict of interest

None to declare.

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Fig. 1. CT scan of the thorax upon admission.