



Spontaneous Bilateral Pneumothorax and Pneumomediastinum Revealing a Bronchial Carcinoid Tumor

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Abstract

Bronchial carcinoid tumors represent 1% to 2% of all pulmonary neoplasias. We report a case where spontaneous bilateral pneumothorax and pneumomediastinum revealed a carcinoid tumor of the right main stem bronchus. KF, 38 years old, exposed to passive smoking for 12 years, was admitted for a spontaneous bilateral pneumothorax and pneumomediastinum. A bronchoscopy has been realized to identify their origin revealing a burgeoning, round, raspberry-tinted and richly vascularized tumor looking like a neuroendocrine tumor at the entry of the right main stem bronchus. That tumor was totally obstructing the bronchus lumen. Bronchial biopsies related an invasive carcinoma few differentiated TTF1. After a negative work-up extension and an operability assessment, a lung-conserving surgery was planned: a resection of the right main stem bronchus and its anastomosis with the upper lobar bronchi and the intermediary trunk with nodes dissection. The surgery was a success permitting to preserve the pulmonary parenchyma.

Lung-conserving strategy in carcinoid tumors treatment should be planned as often as possible permitting to avoid unnecessary pulmonary resections.

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Received Date: 19 Jun 2019

Accepted Date: 02 Aug 2019

Published Date: 12 Aug 2019

Citation:

Ammouri Z, Idelhaj N, Boubia S, Ridai M. Spontaneous

Bilateral Pneumothorax and

Pneumomediastinum Revealing a

Bronchial Carcinoid Tumor. Clin Surg.

2019; 4: 2547.

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Keywords: Bronchial carcinoid tumors; Pneumothorax; Pneumomediastinum; Thoracic surgery; Lung conservative surgery

Introduction

Neuroendocrine tumors of the lung are a heterogeneous group of lung neoplasms. They can be classified into four histological different subgroups that are: Typical Carcinoid (TC), Atypical Carcinoid (AC), Large Cell Neuroendocrine Carcinoma (LCNEC) and Small Cell Lung Carcinoma (SCLC). Bronchial carcinoid tumors (typical and atypical carcinoid) represent 1% to 2% of all pulmonary neoplasias. They are slow-growing tumors and constitute a subgroup among neuroendocrine tumors of the lung. We report a case where spontaneous bilateral pneumothorax and pneumomediastinum revealed a carcinoid tumor of the right main stem bronchus.

Case Presentation

KF, 38 years old, exposed to passive smoking for 12 years, was admitted for thoracic pain and several coughing attacks. Thoracic CT-Scan showed a bilateral low-abundance pneumothorax and a moderate-abundance pneumomediastinum (Figure 1). A bronchoscopy has been realized to identify their origin. It has revealed a burgeoning, round, raspberry-tinted and richly vascularized tumor looking like a neuroendocrine tumor at the entry of the right main stem bronchus. That tumor was totally obstructing the bronchus lumen. Bronchial biopsies related an invasive carcinoma few differentiated TTF1. After a negative work-up extension we started an operability assessment. Spirometry test that showed a decreased Forced expiratory volume in 1 second (FEV1): 35% while diffusing capacity of the lung for carbon monoxide (DLCO) was normal. With these assessments, a lung-conserving surgery was planned: a resection of the right main stem bronchus and its anastomosis with the upper lobar bronchi and the intermediary trunk with nodes dissection. Using a postero-lateral thoracotomy, we managed to resect the tumor which was round, raspberry-tinted and richly vascularized and which had a sessile fastener with bronchial lumen. After the resection, an anastomosis between the origin of the right main stem bronchus with the upper lobar bronchi and the intermediary trunk was made (Figure 2). We ended our surgery with lymphadenectomy among the removed nodes. We had right lower paratracheal (4R), subcarinal and pulmonary ligament. Frozen examinations showed a malignant tumoral proliferation and a healthy bronchial



Figure 1: Bilateral pneumothorax and pneumomediastinum.

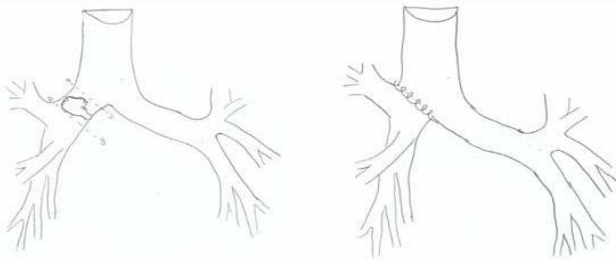


Figure 2: Bronchial tree before and after surgery.

overlap with a reactive adenitis. The surgery was a success permitting to preserve the pulmonary parenchyma.

Discussion

Neuroendocrine Tumors (NETs) are a distinct subgroup of neoplasms arising from the neuroendocrine cells. Due to the peculiar morphological, immunohistochemical and molecular characteristics, NETs are usually classified as a separate group of tumors among solid malignancies.

Pneumothorax is a rare and an exceptional clinical manifestation of lung cancer. It can exceptionally reveal a bronchial carcinoid tumor. Ouede et al. reported a case of a 27-years-old woman in whom recurrent pneumothoraxes were the clinical manifestation of a bronchial carcinoid tumor while the first case ever reported was in 1992 by Wagner et al. [1,2]. In our case, bilateral spontaneous pneumothorax associated with a pneumomediastinum was the clinical manifestation of the bronchial carcinoid tumor. The tumor obstructing the bronchial lumen works like a one-way valve blocking

the inspired air during expiration. This mechanism is at the origin of the pneumothorax and pneumomediastinum and the air blocked inside the lung increases the intrapulmonary pressure leading to the distension of pulmonary alveoli and their rupture in the pleural cavity. This pneumothorax mechanism is totally different from the pneumothoraxes caused by the rupture of an emphysematous bulla or a metastatic lesion [3,4].

Lung-conserving strategy in carcinoid tumors treatment should be planned as often as possible permitting to avoid unnecessary pulmonary resection. Lymphadenectomy should be carried out according to the current guidelines (IASLC/ESTS), including a minimum of 6 nodes (stations) of which 3 mediastinal including the subcarinal one [3]. In our case, the Lymphadenectomy included 3 mediastinal nodes including the subcarinal one and none of them was malignant.

Furthermore, survival after resection is more than 90% excellent for patients with typical carcinoid tumor living up to 10 years and more than 60% for patients with atypical carcinoid tumor.

Conclusion

Bilateral spontaneous pneumothorax and pneumomediastinum are a very rare and uncommon manifestation of bronchial carcinoid tumors. Bronchial carcinoid tumors represent only 1% to 2% of all pulmonary neoplasms.

CT scan and bronchoscopy played a major role in the diagnosis, in the planning of the surgical act and in the prognosis. Lung-conserving strategy in carcinoid tumors treatment should be planned as often as possible permitting to avoid unnecessary pulmonary resections.

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