Case report - Thoracic general

Salivary gland-type mixed tumor (pleomorphic adenoma) of the lung

Angelo Carrettaa,*, Lidia Librettia, Gianluca Taccagnib, Piero Zanninia

aDepartment of Thoracic Surgery, Scientific Institute H San Raffaele, Vita-Salute San Raffaele University, Via Olgettina, 60 -20132 Milano, Italy
bDepartment of Pathology, Scientific Institute H San Raffaele, Vita-Salute San Raffaele University, Via Olgettina, 60-20132 Milano, Italy

Received 31 March 2004; received in revised form 8 July 2004; accepted 23 July 2004

Abstract

Primary pleomorphic adenomas of the respiratory tract are rare tumors. They usually originate from the bronchial glands, but may also be unrelated to the bronchial tract and be localized in the pulmonary parenchyma. These salivary-gland type neoplasms have peculiar histological and clinical features: they usually behave as low-grade malignant neoplasms, but may also have more aggressive features. The diagnostic and therapeutic approach in a patient with a pulmonary pleomorphic adenoma is described.

Keywords: Tumor; Lung; Surgery

1. Introduction

Pleomorphic adenomas of the respiratory tract are rare tumors with peculiar histologic and clinical features [1]. Similarly to their salivary gland counterparts, these neoplasms may either behave as low-grade malignant tumors or have a malignant evolution with a potential to metastatize. Due to the peculiar features of these tumors and to their rarity, several points concerning treatment still need to be defined. We describe the diagnostic and therapeutic approach in a patient with a pulmonary pleomorphic adenoma.

2. Case report

A 22-year-old female patient was admitted to our Department of Thoracic Surgery with a right pulmonary nodule identified during routine preoperative assessment before treatment of an ovarian cyst. Chest X-ray showed a 2-cm rounded lesion, peripherally located in the right pulmonary lower lobe. Chest CT scan showed a well-defined non-calcified nodular lesion of the right lower lobe without enlargement of the hylar and mediastinal lymph nodes (Fig. 1). Resection of the nodule by wedge resection through a right lateral muscle-sparing thoracotomy was performed. Macroscopic examination showed a 2.4-cm yellowish-grey non-capsulated nodule with a smooth surface clearly demarcated from the surrounding lung parenchyma. At histological examination the tumor was mostly organized in cords and nests, with a proliferation of cuboidal epithelial cells without nuclear atypia, forming broad papillae and ductal structures, from small to cystic. Mesenchymal-type cells varied from polygonal to fusiform, with oval and vescicular nuclei, a clear or deeply eosinophilic and fibrillary cytoplasm; myoepithelial cells were also observed. In some areas the cells were organized at the periphery of small ducts. The lesion was also constituted by a variable quantity of stromal/mesenchymal component, mainly formed by round and sometimes confluent islands of chondroid tissue, with large pools of chondroid matrix and dispersed mature chondrocyte cells, sometimes with a clear halo (Fig. 2). At immunohistochemistry proliferating cells were focally PAS positive. Immunoreactivity was positive for cytokeratin, CAM 5.2, EMA, vimentin, S-100 protein, actin, glial fibrillary acid protein and negative for CEA and chromogranin A. Immunocytochemical determination of the MIB-1 proliferative index was lower than 1%. The final diagnosis was salivary gland-type pleomorphic adenoma. Ultrasound evaluation of the salivary glands, performed to exclude a metastatic nature of
the lesion, was normal. Two years after surgery the patient is doing well without tumor recurrence.

3. Discussion

Salivary-gland type tumors of the lung usually originate from the epithelium of the submucosal bronchial gland. They, therefore, usually present as endoluminal lesions and rarely occur in peripheral or subpleural locations [1,2]. The presence of tumors unrelated to the bronchial structure has been explained by a possible origin from a primitive stem cell that can differentiate to a ductal structure, myoepithelium and chondroid or myxoid matrix [3,4].

Three distinct types of pleomorphic adenoma may be recognized: a typical form characterized by a chondromyxoid stroma with a variable quantity of glandular component, a solid variant with a prevalent and pronounced solid myoepithelial growth pattern and a malignant variant that shows marked cellular atypia, frequent mytosis, necrosis and myoepithelial cellular proliferation in a myxoid background. Differential diagnosis with both metastatic lesions from mixed tumors of the salivary glands and other biphasic pulmonary neoplasms such as blastoma, hamartochondroma and carcinosarcoma is mandatory.

The clinical presentation of pleomorphic adenomas of the lung depends on their location. Tumors with endoluminal growth may present with dyspnea, sometimes mimicking bronchial asthma, and hemoptysis. Tumors localized in the lung parenchyma are frequently asymptomatic, although symptoms such as fever, weight loss, pleural effusion and shortness of breath have been reported [1,2]. In patients with endoluminal tumors, the endoscopic evaluation usually shows a lesion with a smooth surface without infiltrative features, frequently with a polypoid shape. CT scan examination usually demonstrates nodular well-defined lesions without infiltration of the surrounding tissues, although features of malignancy may also be observed.

The clinical behavior of pleomorphic adenomas of the respiratory tract is similar to that of salivary gland neoplasms. Small well-circumscribed tumors, without cytological evidence of malignancy usually behave in an indolent fashion whereas less circumscribed tumors with infiltrative features potentially show a tendency to recur and to metastasize. These tumors generally behave as low-grade malignant neoplasms with a long interval between the original diagnosis and local recurrence or distance metastases [1].

Surgery is the treatment of choice for pleomorphic adenomas and of other salivary-gland type tumors of the lung. Long-term follow-up is warranted due to the possibility of a malignant behavior. Standard treatment of patients with advanced disease or tumor recurrence has not been completely defined, although a partial response has been observed after radio- or chemotherapy [2].

In conclusion pleomorphic adenomas of the lung are rare tumors with a potentially malignant behavior. Differential diagnosis with other pulmonary lesions is essential to establish adequate treatment. Long-term follow-up after treatment is mandatory due to the possibility of malignant progression.

References

Appendix A. ICVTS on-line discussion

Author: Dr Sameh Sersar, Mansoura Faculty of Medicine, Cardiotoracic Surgery Department, Mansoura, 123 Egypt

Date: 02-Sep-2004

Message: This is a case of asymptomatic solitary pulmonary nodule. Management of SPN when a preoperative tissue diagnosis is not available remains a controversial issue. Observation for growth, biopsy, and resection are the available options, while aiming to avoid delay in the diagnosis and treatment of lung cancer, false negative results, and resection of benign lesions. Solitary Pulmonary Nodule (SPN) is found in 0.1–0.2% of chest radiographs. Various modalities have been available to aid in the diagnosis of SPN including flexible bronchoscopy, sputum cytology, endobronchial and transbronchial biopsy, computerised tomography (CT) guided percutaneous, fine needle aspiration biopsy (FNAB), helical CT, and Positron Emission Tomographic (PET) scanning [1].

I do like the dictum “watch it or wedge it”. The algorithm described by ponn may be the best way to follow as the odds of malignancy remain indeterminate after an evaluation that may include combinations of imaging modalities and non surgical biopsy which are of paramount clinical prevalence to thoracic surgeons. This odds ratio is the one that should be used to formulate further recommendations and the one quoted to the patient who must ultimately decide how to proceed. It gives the clinicoangiologic suspicion, FNABC and Bronchoscopy an important role and gives the VATS a role in treatment [2].

Poor pulmonary lung function was defined in presence of a FEV1, 1.5 or, 50% predicted. Poor cardiopulmonary status (CPS) was defined as the simultaneous presence of two or more of the following factors: age = 75 years, WHO performance status = 3, poor pulmonary functions (FEV1, 1.5 or, 50% of predicted), co-morbid factors (previous myocardial infarction, current angina, uncontrolled diabetes), poor nutritional status, previous lung resection. The definition of WMWR was of a liberal wedge containing the nodule ensuring at least 2 cm palpable tumor free margins macroscopically free of tumor [3].

Absence of significant lung nodule enhancement ( < or = 15 HU) at CT is strongly predictive of benignity [4].

The enhancing rim sign, seen on chest CT with intravenous contrast enhancement of a solitary pulmonary nodule, has been useful to distinguish some benign pulmonary nodules from malignant pulmonary nodules [5].

References: