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Case report

Pneumocytic adenomyoepithelioma: A case providing support for the benignity of this extremely rare pulmonary neoplasm

Brandon T. Nokes[a,*], Anjuli M. Brighton[b], Brandon T. Larsen[c], Prasad M. Panse[d], Staci E. Beamer[e], Rodrigo Cartin-Ceba[b], Karen L. Swanson[b]

[a] Department of Internal Medicine, Mayo Clinic, Phoenix, AZ, USA
[b] Division of Pulmonary Medicine, Mayo Clinic, Phoenix, AZ, USA
[c] Department of Laboratory Medicine and Pathology, Mayo Clinic, Phoenix, AZ, USA
[d] Department of Radiology, Mayo Clinic, Phoenix, AZ, USA
[e] Department of Cardiothoracic Surgery, Mayo Clinic, Phoenix, AZ, USA

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ABSTRACT

Pneumocytic adenomyoepithelioma is an extremely rare and poorly understood pulmonary neoplasm, so experience with this tumor is limited. Since the initial case series where the lesion was first proposed as a distinctive entity, only one additional report has been described. We present a case of pneumocytic adenomyoepithelioma with clinical and radiologic data that provide the first long-term evidence of the benignity of this extremely rare pulmonary neoplasm. We also review the available literature surrounding pneumocytic adenomyoepitheliomas. Our case provides important new data on the behavior of this lesion, as imaging studies showed essentially stable or very slowly progressive disease over the course of approximately 9 years. Collectively, this rare and poorly described lesion appears to behave in an indolent or benign fashion, a notion that our case further supports.

1. Introduction

Benign pulmonary neoplasms are a heterogeneous and often incidentally discovered group of disparate lesions with highly variable prognostic implications [1]. Among these lesions is a group of rare neoplasms showing both epithelial and myoepithelial differentiation, most likely arising from minor salivary gland cells within the large airways of the lower respiratory tract, representing the pulmonary analogues of their far more common counterparts in the major salivary glands of the head and neck [2]. Although this family of tumors has been recognized for many years and most have been relatively well characterized, in 2007 Chang et al. described 5 cases of a distinctive and previously unrecognized pulmonary neoplasm showing not only epithelial and myoepithelial differentiation, but also evidence of pneumocytic differentiation, which they termed pneumocytic adenomyoepithelioma [3]. These tumors are extraordinarily rare and their clinical presentation and behavior remain poorly understood. Although it has been suggested that they behave in an indolent or benign fashion, no long-term clinical or radiologic evidence has been reported to support this notion and published data on this lesion are limited. Here, we present a case of pneumocytic adenomyoepithelioma with clinical and radiologic data that provide the first long-term evidence of the benignity of this extremely rare pulmonary neoplasm. We also review the available literature surrounding pneumocytic adenomyoepitheliomas.

2. Case presentation

A 63-year-old woman presented to our pulmonary clinic for evaluation of multiple bilateral pulmonary nodules. She had been recently hospitalized for a transient abdominal illness at an outside hospital, and an abdominal CT at that time detected multiple subcentimeter lesions in the bilateral lower lobes, but was otherwise unremarkable. Her past medical history was notable for hypertension, diabetes, gastroesophageal reflux, thalassemia, and remote and treated pulmonary coccidioidomycosis, which was previously identified by serology without associated x-ray findings at the time of diagnosis. She was a lifelong nonsmoker and did not have any known occupational exposures, but endorsed a passive smoke exposure in her home environment and had a family history of lung cancer.

She had also been seen in our pulmonary clinic eight years prior for incidentally detected bilateral pulmonary nodules. These nodules were initially detected in February 2008 at an outside facility. On our

* Corresponding author. Department of Internal Medicine, Mayo Clinic, 13400 East Shea Boulevard, Scottsdale, AZ, 85259, USA.
E-mail address: Nokes.Brandon@mayo.edu (B.T. Nokes).

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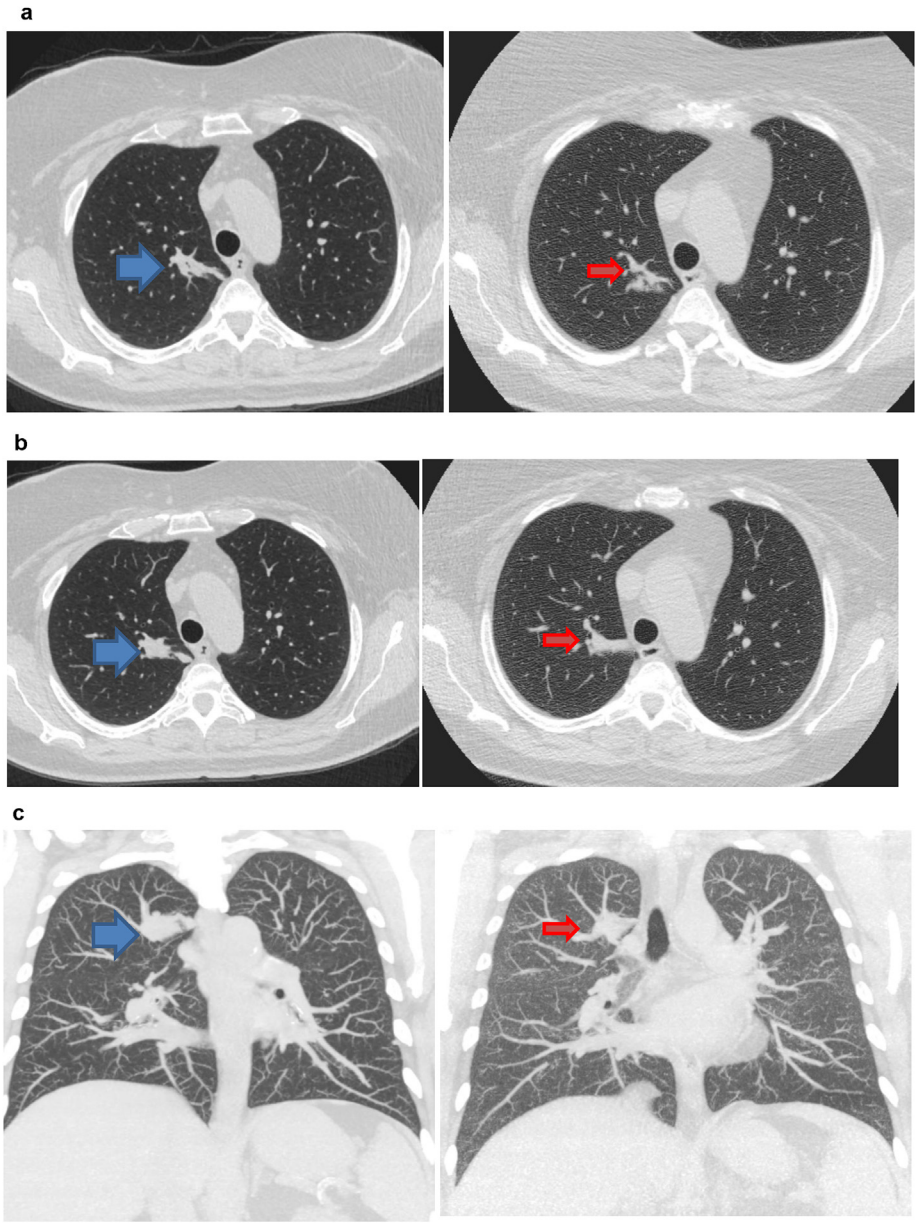

Fig. 1. a. Nodule in the right upper lobe centered around the bronchus (solid blue arrow) on September 2016 slowly growing since December 2008 (red arrow). **b**. Axial CT image slightly inferiorly shows the slowly enlarging nodule on September 2016 (solid blue arrow) as compared with December 2008 (red arrow). **c**. Coronal multiplanar reconstruction CT image shows the slowly enlarging nodule on September 2016 (solid blue arrow) as compared with December 2008 (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

assessment in May 2008, a chest CT without contrast showed findings radiologically consistent with several benign intrapulmonary lymph nodes and subpleural lymph nodes in the posterior portion of both lower lobes. Benign-appearing linear fibrosis was also noted in the right upper lobe apical segment medially and in the inferior lingular segment. In the right upper lobe, this abnormality was possibly associated with an endobronchial lesion (Fig. 1a), although this was not well appreciated at the time of initial evaluation and this was only recognized on subsequent review many years later when she presented again. Follow-up imaging 7 months later in 2008 was unchanged and ultimately these findings were felt to be benign without the need for subsequent imaging.

On our imaging assessment in 2016, a CT scan without contrast demonstrated a dominant slowly growing 14 mm nodule apparently centered along a subsegmental branch of the right apical segmental bronchus (Fig. 1b), in the same area where linear fibrosis was noted in 2008. The nodule appeared to be associated with airway obstruction and mucous plugging. Given the extremely slow interval growth and the fact that the lesion was centered on an airway, a typical carcinoid tumor was suspected. In addition, the patient had increasing mediastinal lymphadenopathy, predominantly in the right paratracheal region. Multiple additional subcentimeter pulmonary nodules were noted, many of which appeared to have a centrilobular distribution. Some of these nodules were stable back to 2008 and some had slightly increased in size. Additionally, there were numerous indeterminate nodules within the lungs bilaterally, as well as some new nodules along the fissures, which were new since December 2008.

Multiple prominent mediastinal lymph nodes were also noted as well as multiple indeterminate nodules along the fissures (Fig. 1c). Accordingly, bronchoscopy was pursued.

At the time of bronchoscopy, using the BCUC180F flexible fiberoptic bronchoscope, an endobronchial lesion was visualized in the right