A rare case of primary sclerosing pneumocytoma in a healthy female

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Introduction

Pulmonary sclerosing pneumocytoma is a rare group of benign pulmonary neoplasms that usually affects older adults over 50 years of age [1]. This neoplasm is more commonly seen in women, with female to male ratio of 5:1 and is predominantly seen in Asian ethnicities [2]. These tumors originate from type II alveolar pneumocytes and are hence classified as adenoma under the 2021 World Health Organization (WHO) classification of tumors [3]. They account for 3-5 percent of total benign lung tumors [4].

One of the striking aspects of PSP is its clinical presentation, or rather the lack thereof in many cases. It is often discovered incidentally during routine chest imaging, such as chest X-rays or CT scans, due to its asymptomatic nature. However, when symptoms do occur, they tend to be non-specific, including cough, chest pain, or respiratory distress. This variability in clinical manifestation further complicates the diagnosis of PSP, as these symptoms overlap with those of other pulmonary conditions. Moreover, it is essential to consider the potential for malignant transformation, as a subset of PSP cases have exhibited aggressive behavior [5].

Herein, we describe a case of a 73-year-old female who was found to have an incidental lung nodule. In view of the history of interval increase in the size of the lung nodule, the patient underwent a CT-guided lung biopsy, which was characterized as sclerosing pneumocytoma, and the patient was referred to thoracic surgery for subsegmental right middle lobe resection.

Case History

A 73-year-old female was seen in her cardiologist’s office for a routine visit. The patient had a past medical history significant for hypertension, hyperlipidemia, and coronary artery disease diagnosed on CT imaging. The patient was incidentally also found to have a lung nodule in her right middle lobe. In the office, the patient was completely asymptomatic; a review of systems was negative for any chest pain, cough, shortness of breath, recent weight loss, loss of appetite, fevers, or night sweats. The physical exam was within normal limits. The patient has no history of smoking nicotine or any other drugs, worked as a chef in the food service industry, and denies any occupational metal/fumes exposures.

Methods

Investigations:

CT imaging showed marginated, noncalcified pulmonary nodule in the right middle lobe measuring 2.6*2.4 cm (Figure 1A). There was no evidence of hilar/mediastinal lymphadenopathy, other nodules, septal thickening, or interstitial lung disease. The patient had a chest CT 4 years prior, on which this nodule
measured 1.6*1.6 cm. The patient was referred to pulmonologist and was recommended for a PET scan, which revealed a 2.8*2.9 cm well-circumscribed mildly hypermetabolic lesion in the with SUV of 2.68 and tissue sampling was recommended (Figure 1B, 1C). Of note, no other hypermetabolic lesions were found anywhere else in the body. Thereafter, the patient underwent bronchoscopy with broncholoalveolar lavage and a transbronchial biopsy. The biopsy only showed hematoxyn, and BAL cytology was negative. However, due to concern for interval growth of the nodule and hypermetabolic PET, a CT-guided biopsy was pursued. Pathology revealed pulmonary sclerosing neurocytoma with hematoxylin and eosin staining showing classic cuboidal surface cells and round stromal cells (Figure 2). Immunohistochemistry was positive for AE 13, TTF-1 and EMA; the tumor was negative for Napsin and ERG (Figure 3).

**Differential diagnosis**

Radiographically and pathologically, PSP can mimic multiple benign, primary, and metastatic neoplasms, namely clear cell tumors, hamartomas, hemangiomas, carcinoids and bronchoalveolar carcinoma. Immunohistochemistry is the cornerstone for confirming the diagnosis [5, 6, 7].

**Treatment course**

The patient was referred to see oncology and thoracic surgery. The patient’s case was discussed at thoracic tumor board and was recommended video assisted thoracic surgery guided right middle lobectomy.

**Conclusion and Results**

The patient opted for active surveillance instead, with repeat imaging in 4 months and regular flow up with oncology, pulmonology, and surgery.

Pulmonary sclerosing pneumocytoma is a captivating and rare pulmonary tumor that remains a diagnostic challenge due to its varied clinical presentation and distinctive histopathological features. While most cases are benign and amenable to surgical treatment, the potential for malignant transformation underscores the importance of accurate diagnosis and long-term monitoring.

**Discussion**

Pulmonary sclerosing pneumocytoma is most commonly present as an asymptomatic finding and is usually incidentally diagnosed on imaging done for unrelated reasons. In less than 10% of patients, it can present with nonspecific symptoms such as cough, chest pain, and shortness of breath [5]. Although benign, these tumors have high potential for proliferation, and the size of the tumor is highly variable, ranging from a subcentimeter to up to 12 cm [8]. They are also associated with local recurrence and have been known to metastasize to lymph nodes, pleura, and bones [8]. Radiographically and morphologically, neurocytomas are usually well-circumscribed, homogenously enhanced, and unencapsulated [9]. Pathologically, they are composed of two cell types- cuboidal pneumocytes and round or polygonal stromal cells, and four different patterns have been described, namely papillary, solid, hemorrhagic, and sclerotic [10].

The most common immunohistochemical markers are EMA and TTF-1[11]. These tumors also tend to be progesterone receptor positive hence the female predominance. Molecular alterations that have been most commonly described include mutations in the AKT-1, beta-catenin, BRAF genes [12] and [13]. A neurocytoma harboring p53 and PI 3 kinase mutations was reported in Shanghai, China. This tumor was associated with multifocal lymph node metastasis. [14]

The prognosis of PSP is excellent due to its benign nature. Out of 3496 patients reported till date there is only one cancer related death due to an extensive disease burden causing respiratory failure [15]. First-line treatment modality is usually surgical resection. Both lobectomies and sub-lobectomies have been performed, depending on the tumor burden Sub-lobectomies including segmentectomy and wedge resections have shown better tolerability and patient outcomes, including shorter length of hospital stay, low risk of postoperative complications as compared to lobectomy [2]. Interestingly, in a retrospective study of 107 PSP patients in South Korea, no difference was observed in all-cause mortality between surgical management and active surveillance [16].
Future directions

The current standard of care is surgical resection; however, in cases of future recurrence and metastasis, molecular profiling can be considered to treat with targeted immunotherapy agents. Since AKT-1 has been found to be the driver mutation in most of the PSC cases, its inhibitors theoretically should help. No studies have been done so far since this entity is so rare and metastasis is even more remote [17].

References


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Figures

Figure 1- A- 2.8 x 2.9 cm well-circumscribed nodule with a subtle component of calcification is noted in the right middle lobe.

B, C- right middle lobe nodule with mild increased metabolic activity with max SUV of 2.68 in axial and coronal planes.

Figure 2- Hematoxylin Eosin stain

Pathological findings of Pulmonary sclerosing pneumocytoma showing 2 types of cells, cuboidal surface cells and stromal round cells. A: Low power, B: High power

Figure 3- Immunohistochemistry profile

A-AE1/3 positive for surface cells, B-EMA positive for both cell population, C-TTF-1 positive for both cell population

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