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Case Report

Treatment and post-operative follow-up of pulmonary sclerosing pneumocytoma: A case report

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ABSTRACT

Introduction and importance: Pulmonary sclerosing pneumocytoma (PSP) is a rare tumor thought to originate from respiratory epithelial cells. It is usually benign, but may rarely metastasize to lymph nodes. Surgeons face unique challenges in diagnosis and management of this condition, and ideal surgical management is yet to be established.

Case presentation: 48-year-old woman with a 7 × 7 mm pulmonary lesion discovered incidentally on computerized tomography (CT) imaging, which grew to 9 mm over the following year. Seven years later, follow-up imaging revealed that the mass had grown to 1.3 cm in largest dimension. Surgery was recommended and the mass was resected via a right video-assisted thoracic surgery (VATS) middle lobectomy with mediastinal lymph node dissection. All lymph nodes were negative and the patient's postoperative course was unremarkable.

Clinical discussion: There are few evidence-based guidelines available on the treatment and postoperative surveillance of PSP. Research has shown comparable recurrence-free survival rates for sublobar resection and lobectomy, though recurrence can occur, especially following sublobar resection in larger or more centrally-located tumors. In absence of established guidelines, it was decided to follow this patient according to NCCN guidelines for surveillance of early-stage non-small cell lung cancer due to potential risk of recurrence.

Conclusion: This case report adds to the limited literature on PSP and depicts a possible treatment and post-operative follow-up plan. Right VATS middle lobectomy can effectively treat some cases of central PSP. In absence of established guidelines for postoperative follow-up of PSP, NCCN guidelines may outline one possible strategy for postoperative management.

1. Introduction and importance

Pulmonary sclerosing pneumocytoma (PSP), formerly known as sclerosing hemangioma of the lung, is a rare tumor of the lung first described by Leibow and Hubbell in 1956 [1,2]. Here we present a 48-year old woman with incidentally-discovered PSP treated with a right video-assisted thoracic surgery (VATS) middle lobectomy and mediastinal lymph node dissection (MLND) and review the literature surrounding this rare diagnosis. This report also describes a post-operative follow-up imaging plan given the absence of published guidelines for recurrence surveillance after resection of PSP. This work is reported in line with the Surgical Case Report (SCARE) criteria [3].

PSP originates from the respiratory epithelium and is most likely derived from type II pneumocytes [1,4,5]. PSP is generally asymptomatic [6] and is usually benign in nature, but carries the potential to have malignant characteristics such as lymph node metastasis or local recurrence [1,7,8]. Additionally, this rare mass is frequently mistaken for lung cancer malignancies upon initial imaging due to diagnostic difficulties, as many of the signs seen on imaging are non-specific [9,10].

Given the relatively benign course of PSP and its low prevalence, therapeutic management has not yet been definitively established. Surgical resection, including enucleation, lobectomy, or sublobar

Abbreviations: PSP, pulmonary sclerosing pneumocytoma; VATS, video-assisted thoracic surgery; CT, computerized tomography; PET, positron emission tomography; SUV, standardized uptake value.

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resection (wedge, segmentectomy), is the only known curative treatment course [8,11]. Surgical technique may vary based on the size and location of the tumor [11].

2. Case presentation

A 48-year-old woman who is a never-smoker of Asian descent initially presented nine years previously with unrelated lower abdominal pain. The patient had no relevant past medical or surgical history, and no relevant family history. Computerized tomography (CT) scanning of the abdomen and pelvis revealed an incidental finding of a round 7 × 7 mm nodule in the right middle lobe of the lung. The patient had no pulmonary symptoms and the remainder of her physical exam and labs were unremarkable. Subsequent CT imaging of the thorax approximately two months later confirmed a 7 × 8 × 6 mm non-calcified pulmonary nodule in the right middle lobe of the lung. Lungs appeared clear otherwise.

Following the Fleischner Society recommendations for low risk patients [12], this patient received a follow-up CT scan one year later which revealed an interval increase in size of the right middle lobe lung nodule to 9 mm in largest dimension. CT imaging of the thorax was performed again six months later and revealed no further change in size of the nodule. At this time, pulmonology recommended resection given the tumor growth over the previous year. However, the patient chose to not be referred to thoracic surgery at that time and was lost to follow-up.

Seven years later, the patient underwent another follow-up CT scan of the chest which revealed substantial growth of the non-calcified right middle lobe pulmonary nodule to 13 mm in largest dimension (Fig. 1). Positron emission tomography (PET) imaging showed minimal tracer avidity of the pulmonary nodule (SUV 1.8). No concerning mediastinal lymph nodes were noted.

At this time, the patient consented to surgery. Multiple surgical approaches were considered and discussed with the patient preoperatively, including right VATS sublobar resection of the right middle lobe if feasible or straight to right VATS middle lobectomy with MLND depending on intraoperative assessment and location. Due to the size and central location of the nodule, a right VATS middle lobectomy with MLND was performed by an attending thoracic surgeon at a regional general hospital. Lymph nodes were sampled from levels 2, 4, 7, 10, 11, 12, and 14. Upon pathology, the nodule was confirmed to be a right middle lobe sclerosing pneumocytoma measuring 1.3 × 1.2 × 0.8 cm, and pathological staging was T1bN0M0. All margins were negative.

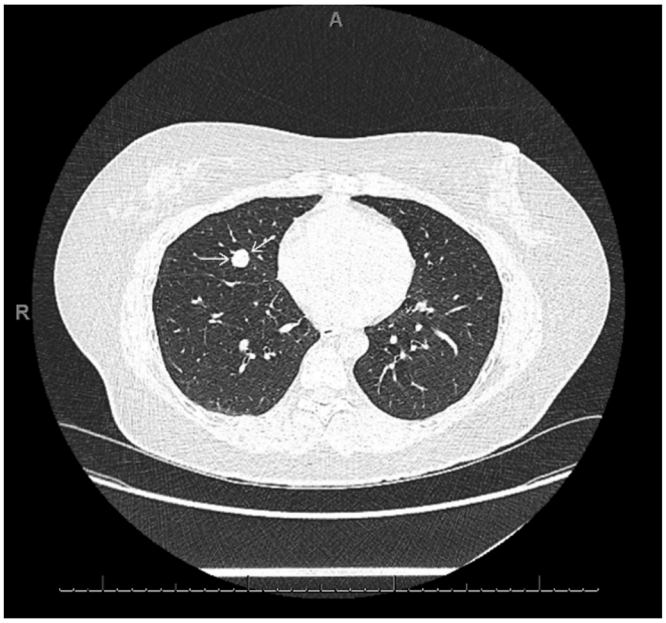

Fig. 1. CT Scan. *1.3 × 1.2 × 0.*8 cm *pulmonary nodule of the right middle lobe.*

Her post-operative course was uneventful. She was discharged the following day. At this time, she was tolerating a regular diet, ambulating, and had adequate pain control on oral medications. This patient will receive follow-up CT chest imaging to assess for recurrence.

3. Clinical discussion

PSP is a rare pulmonary tumor most commonly occurring in adults over 50 years of age [13], and is significantly more common in women, especially of Asian descent [4,13]. It is most frequently asymptomatic and discovered incidentally via imaging [9], though symptoms can include cough and chest pain [14]. Rarely can these tumors lead to hemoptysis or airway obstruction [14].

Diagnostic challenges in PSP stem from non-specific imaging findings that can commonly be misdiagnosed as primary or metastatic lung malignancies [9]. On CT imaging, PSP is usually found as a well-defined single nodule, often with surrounding ground-glass opacity [9]. Only a minority of patients with PSP are correctly diagnosed via conventional CT imaging [9]. Immunohistochemistry is not done in every case, but can help differentiate PSP, which generally present as hypometabolic nodules, from pulmonary malignancies, which are hypermetabolic [9]. Pathology can confirm the diagnosis of PSP and further discriminate from lung adenocarcinoma. On pathology, PSP is identified via cuboidal surface cells and stromal round cells [10], both of which are positive for thyroid transcription factor-1 [15].

Once PSP has been accurately diagnosed, therapeutic challenges of this rare tumor remain. Surgical resection is generally considered to be the only curative treatment. However, the optimal operative approach has been debated [8]. Enucleation is a common approach, followed by lobectomy or sublobar resection [11]. Lymph node dissection may be done to assess for metastasis of PSP and to rule out non-small cell lung cancer [7,8]. Radiotherapy has been suggested as an alternative for patients with inoperable tumors [16].

Currently, lobectomy is the surgical approach used most frequently in patients with centrally-located tumors near the hilum of the lung [11]. A previous study by Park et al. [17] showed that all patients undergoing either lobectomy or sublobar resection of PSP were free of local recurrence or distant metastasis throughout the entire follow-up period, andpatients who underwent limited resection group had a significantly shorter mean hospital stay in comparison to patients who underwent lobectomy [17]. However, other reports have described rare recurrence of PSP after sublobar resection [18]. Adjuvant therapy is generally not considered due to the benign nature of this disease and low recurrence rates [4,6–8,11].

At this time, it appears that a minimally invasive sublobar resection is an appropriate surgical approach for small, peripherally-located PSPs, while minimally invasive lobectomy should be the choice approach for larger tumors or those that are more centrally-located with greater potential for lymph node metastasis or recurrence [1]. However, further research in larger cohorts comparing the two approaches longitudinally are needed to clearly define these recommendations.

Although recurrence of PSP after resection is rare, there have been several previous reports describing malignant spread of PSP to regional lymph nodes [7,8] as well as local recurrence [18]. Thus, we propose that post-resection surveillance of these patients is justified. There are no current recommendations to guide post-operative follow-up after PSP resection, so it was elected to perform follow-up imaging in accordance with current NCCN Guidelines for early stage non-small cell lung cancer in this patient [19]. Therefore, this patient will receive chest CT imaging every six months for at least two years, and then low-dose non-contrast enhanced chest CT scans annually after completion of definitive therapy [19]. There is a need to establish evidence-based recommendations for post-resection surveillance of patients with PSP to ensure a standardized approach to care.

This case report represents a single incidence of PSP, and therefore is not sufficient to guide treatment in all cases. Additionally, the patient has yet to begin her postoperative surveillance via chest CT imaging, so adherence to the postoperative follow-up has not yet been assessed. Nevertheless, this case report adds to the limited literature on PSP and may help elucidate certain considerations that can affect treatment choice and the decision of whether to and how to conduct postoperative follow-up imaging in cases of PSP.

4. Conclusion

Here we present a patient with a 1.3 cm central PSP of the right middle lobe of the lung that was treated via right VATS middle lobectomy with MLND. This patient will receive follow-up imaging in accordance with current NCCN Guidelines for early-stage non-small cell lung cancer due to the potential risk of recurrence. The purpose of this report is to add to the limited literature on PSP and highlight that right VATS middle lobectomy with MLND is a safe option for treatment of these rare tumors. This report also details a potential postoperative follow-up plan in consideration of the absence of published universal guidelines available on frequency of follow-up imaging for PSP.

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Author contribution

Katherine E. Barnes: data curation, writing – original draft and editing;
Rachel K. Wile: data curation, writing – original draft and editing;
Kian C. Banks: conceptualization, writing – review and editing, supervision;
Jeffrey B. Velotta: conceptualization, writing – review and editing, supervision.

Registration of research studies

Name of the registry: n/a
Unique Identifying number or registration ID: n/a
Hyperlink to your specific registration (must be publicly accessible and will be checked): n/a

Guarantor

Jeffrey B. Velotta.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104836.

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