Letter to the Editor

Pulmonary sclerosing pneumocytoma with rapid progression: a case description of serial images

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Submitted Sep 16, 2024. Accepted for publication Jul 24, 2025. Published online Oct 22, 2025.
doi: 10.21037/qims-24-1930
View this article at: https://dx.doi.org/10.21037/qims-24-1930

Introduction

Pulmonary sclerosing pneumocytoma (PSP) is a rare benign neoplasm primarily observed in the middle-aged female demographic (1,2). Characteristically indolent, PSP is typically asymptomatic in most patients. Notwithstanding, a subset of patients may present with non-specific respiratory symptoms, including cough, expectoration, and hemoptysis, as well as discomfort that manifests as chest or back pain (3). Histologically, PSP has relatively innocuous cytological features and a propensity for slow proliferation (4). Here, we present an atypical instance of PSP characterized by rapid progression, and outline possible computed tomography (CT) hallmarks of the disease, thereby enhancing the understanding of this disease entity.

Case presentation

A 50-year-old non-smoking female patient without significant medical or family history was incidentally found to have a solitary lung nodule on low-dose chest CT during a routine annual health check-up. The nodule, located in the left lower lung lobe, measuring 7.9 mm × 7.9 mm with homogeneous attenuation, was presumed to be benign, and was initially managed conservatively with surveillance. Over time, however, the nodule gradually enlarged, measuring 11.8 mm × 10.4 mm after two years and 17.2 mm × 16.8 mm at the 5-year follow-up (*Figure 1*). The growth was accompanied by the appearance of surrounding ground-glass opacity (GGO), prompting the patient to seek further medical evaluation. Upon calculation, the nodule's doubling time was estimated to be approximately 426 days. At the time of presentation, the patient was asymptomatic, with no fever, chest tightness, chest pain, hemoptysis, or night sweats.

The patient was admitted for a comprehensive examination. Laboratory investigations, including complete blood count, cardiac enzymes, coagulation profile, erythrocyte sedimentation rate (ESR), tumor markers, and a 7-autoantibody panel for lung cancer, were unremarkable. Contrast-enhanced CT demonstrated interval enlargement of the left lower lobe nodule with extensive surrounding GGOs (*Figure 2*). 18F-fluorodeoxyglucose positron emission tomography-CT (^{18}F-FDG PET/CT) revealed a mildly hypermetabolic lesion [maximum standardized uptake value (SUV_{max}) 1.4] exhibiting smooth margins and no associated lymphadenopathy.

A definitive diagnosis was established via left lower lobectomy with systematic lymph node dissection under intercostal nerve block anesthesia. Histopathological examination of the resected specimen confirmed PSP

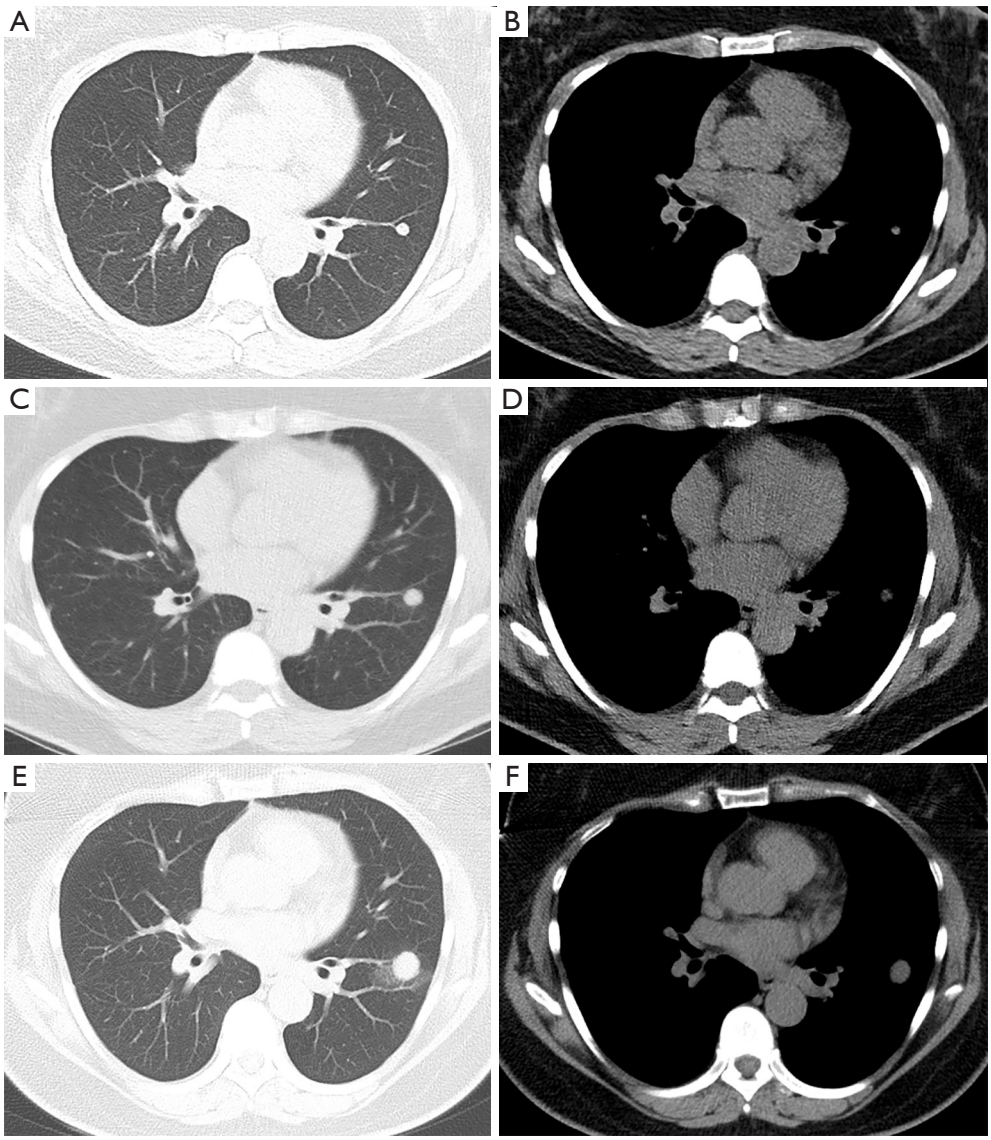

Figure 1 Serial CT images obtained in a 50-year-old woman with PSP. CT images of a solitary pulmonary nodule diagnosed as PSP in a 50-year-old woman showing progressive increase in size over time. Initial axial CT images of the left lower lobe (A, lung window; B, mediastinal window) reveal a nodule measuring 7.9 mm × 7.9 mm with homogeneous texture. Follow-up imaging after two years demonstrates enlargement of the nodule to 11.8 mm × 10.4 mm (depicted in subsequent lung and mediastinal windows, C and D). After another three years, the nodule has further increased to 17.2 mm × 16.8 mm, accompanied by the appearance of surrounding GGO (E, lung window; F, mediastinal window). CT, computed tomography; GGO, ground-glass opacity; PSP, pulmonary sclerosing pneumocytoma.

(*Figure 3A*). Immunohistochemical profiling supported this diagnosis: the lesion showed negative reactivity for ALK-D5F3, P40, P63, Desmin, Syn, S-100, and SMA, yet demonstrated positive staining for epithelial markers [cytokeratin 7 (CK7), Napsin A, thyroid transcription factor-1 (TTF-1)], as well as vascular markers (CD31, CD34, and ERG) (*Figure 3B,3C*).

This study was approved by the Institutional Review Board of Peking Union Medical College Hospital and conducted in accordance with the Declaration of Helsinki and its subsequent amendments. Written informed consent was obtained from the patient for publication of this article and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

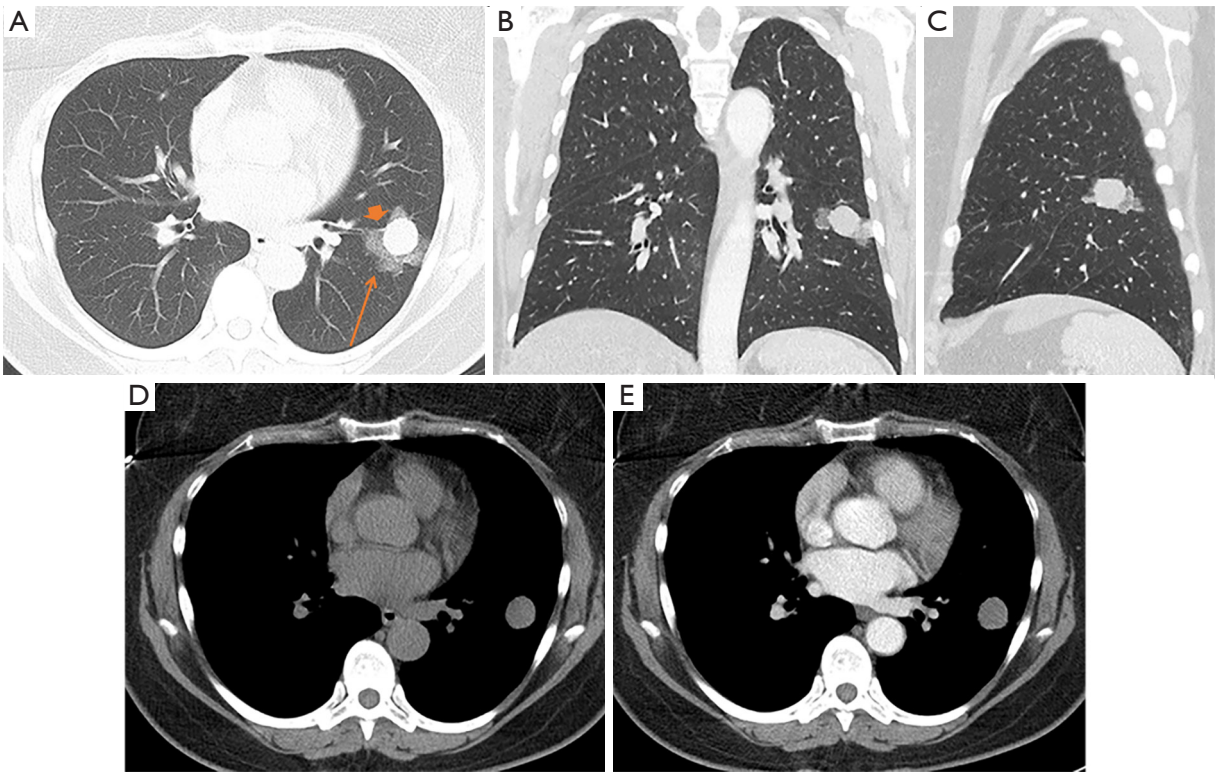

Figure 2 Preoperative contrast-enhanced CT scans in a 50-year-old woman with PSP. (A-C) On the lung window, a solitary nodule in the left lower lobe measuring 21.9 mm × 19.8 mm was noted, with surrounding GGO, that is, "halo sign" [the long arrow in (A)]. A linear projection is observed emanating from the lesion, that is, "tail sign" [the short arrow in (A)] (A, axial; B, coronal; C, sagittal). (D,E) A slight homogeneous enhancement of the nodule is evident upon contrast administration (D, unenhanced mediastinal window image; E, post-contrast mediastinal window image). CT, computed tomography; GGO, ground-glass opacity; PSP, pulmonary sclerosing pneumocytoma.

Discussion

PSP is a rare pulmonary tumor, typically exhibiting a benign clinical progression. Histologically complex, it exhibits four architectural patterns: solid, papillary, sclerotic, and hemorrhagic, composed of dual cell populations that are phenotypically similar to type II pneumocytes and round stromal cells (2,4). PSP most commonly occurs in middle-aged adults, with a median age of presentation of 50 years, and has a notable gender disparity, with females experiencing a 5:1 risk ratio compared to males (3). Clinical manifestations are often inconspicuous, leading to the disease's frequent incidental discovery during routine examinations. Nevertheless, a minority of patients may present with non-specific respiratory symptoms, such as cough, expectoration, and hemoptysis, as well as localized thoracic or dorsal pain (5). In this case, the patient was asymptomatic, and the pulmonary nodule was incidentally discovered during a routine health screening. Given this unusual radiographic progression, the case was reviewed by a multidisciplinary tumor board. This highlights the importance of considering not only malignancies but also benign neoplasms, particularly those exhibiting atypical behavior, in such multidisciplinary settings.

PSP demonstrates distinctive imaging features on CT. Typically, it presents as a solitary, slow-growing, well-demarcated nodule with smooth margins and homogeneous attenuation on non-contrast scans. Post-contrast imaging reveals intense homogeneous enhancement, reflecting its consistent internal vasculature. Another notable feature is the "halo sign", which manifests as GGO encircling the lesion and reflects hemorrhages arising from the differential contractions between the tumor's fibrous capsule and the neoplastic cells (6). Vital imaging markers for PSP include its dynamic expansion, slow growth, and the intensified density of the surrounding GGO, all of which hint at its

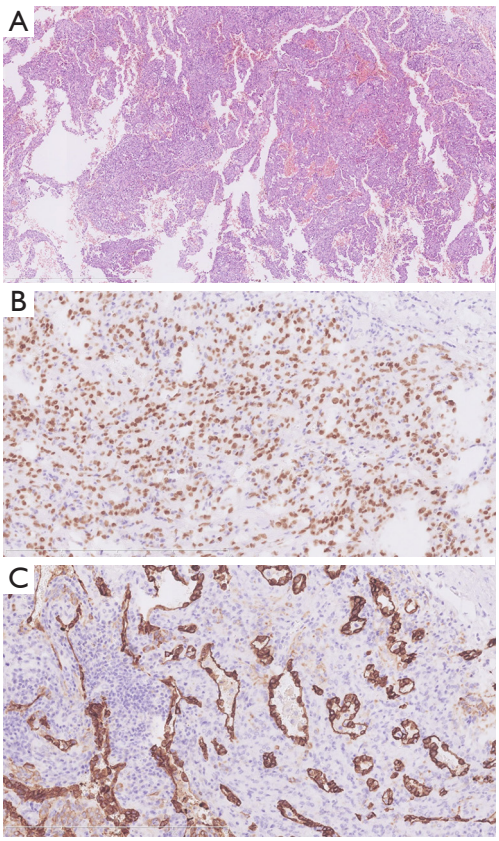

Figure 3 Histopathological and immunohistochemical features of PSP. (A) H&E staining (40× magnification): reveals a biphasic tumor architecture composed of papillary regions and solid regions. The papillary region is predominantly composed of cuboidal surface cells intermingled with rounded cells, whereas the solid region shows rounded cells of uniform size. In the hemorrhagic area, a multitude of red blood cells are observed within the lung interstitium and alveolar spaces. (B) Immunohistochemistry for Pan-CK (160× magnification): demonstrates positive immunoreactivity in cuboidal surface cells, with rounded cells showing no staining. (C) Immunohistochemistry for TTF-1 (160× magnification): shows positive expression in the majority of tumor cells. H&E, hematoxylin and eosin; Pan-CK, pan-cytokeratin; PSP, pulmonary sclerosing pneumocytoma; TTF-1, thyroid transcription factor-1.

non-encapsulated, lepidic growth (7). Tumor infiltration into neighboring lung tissue can also present as GGO (8). For our case, as the size of the lesion increased, so did the size and density of the surrounding GGO, indicating disease progression. Our patient also had the "tail sign", characterized by a diminutive linear extension emerging from the lesion's surface, which is another common CT finding in PSP (9). Meanwhile, the diagnostic imaging features of PSP (7) include the marginal pseudocapsule sign, indicative of compressed lung tissue adjacent to the tumor, and the overlying vessels sign, reflecting pulmonary vessels traversing the tumor surface. The air gap sign, resulting from alveolar mesenchymal cell changes, along with cystic-like alterations and coarse spotty calcifications suggestive of cholesterol clefts, can significantly support a PSP diagnosis. These imaging characteristics are crucial for differentiating PSP from other pulmonary nodules.

The imaging features of PSP overlap significantly with those of lung adenocarcinoma, posing diagnostic challenges. Previous studies have suggested that dynamic contrast-enhanced CT (DCE-CT) demonstrates utility in differentiation (5). PSP typically exhibits early intense enhancement with prolonged retention, correlating histologically with vascular-rich hemangiomatous components. Using radiomics or texture features from CT scans can thus help to differentiate between PSP and atypical peripheral lung cancer (9,10). ^{18}F-FDG PET/CT can also help to differentiate PSP from other conditions, although tumor size and other clinical characteristics may affect FDG uptake in cases of PSP (11). PET/CT images showing round or ovoid lesions with smooth margins and mild to moderate uptake, particularly in middle-aged women, can indicate PSP (12).

Surgical resection constitutes the definitive management for PSP (13). Although PSP is generally regarded as benign, its growth kinetics warrant careful consideration. Prior research (2) suggested that PSPs grow at a rate of 660–1,250 days, which is slower than that of malignant tumors. Notably, our case exhibited accelerated growth with a doubling time of 426 days. Additionally, a recent study reported on PSPs that showed wax-and-wane patterns of growth, including instances where internal hemorrhages or resorption altered tumor size, and times when PSP concurrently presented with atypical hyperplasia, suggesting an inherent potential for growth (14). These dynamics may explain the progressive peri-nodular GGO observed in our patient, where GGO expansion paralleled tumor growth.

Despite the prevailing belief that PSP is a benign condition, some clinicians contend that it represents a potentially malignant or low-grade tumor due to its proclivity for aggressive behaviors. These include lymph node metastasis (15,16), distant or pleural metastasis (17-19), recurrence (20), malignant transformation (21), and death (22). Nevertheless, a substantial body of evidence shows favorable outcomes after surgical excision of localized

recurrences. Moreover, the overall mortality risk specifically attributable to PSP, regardless of surgical intervention, has not been shown to impact overall mortality rates (1). From a histological perspective, positive immunohistochemistry results for epithelial membrane antigen (EMA), TTF-1, and CK-7 indicate that PSP likely originates from pulmonary epithelial cells. Furthermore, somatic mutations in the *AKT1* gene are implicated in PSP pathogenesis (23,24), as they drive abnormal signaling pathways that enhance oncogenic properties such as cell proliferation, survival, and tumor formation (25).

Conclusions

We have reported a case of rapidly progressing PSP. Imaging signs, including the halo sign, tail sign, marginal pseudocapsule sign, overlying/overlapping vessel sign, and air gap sign, may aid in PSP diagnosis, although biopsy remains the gold standard. Short-term follow-up is also essential for cases that are challenging to diagnose. Additionally, DCE-CT, FDG PET/CT, and emerging machine learning models hold promise for assisting in the diagnosis and management of PSP in clinical practice.

Acknowledgments

None.

Footnote

Funding: This work was supported by National High Level Hospital Clinical Research Funding (Nos. 2022-PUMCH-B-069, and 2022-PUMCH-A-034) and Peking Union Medical College Hospital Research Funding for Postdoc (No. kyfyjj202412).

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-24-1930/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This study was approved by the Institutional Review Board of Peking Union Medical College Hospital and conducted in accordance with the Declaration of Helsinki and its subsequent amendments. Written informed consent was obtained from the patient for publication of this article and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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Cite this article as: Piao S, Ming Y, Zhang Y, Zhang H, Zhao R, Wang J, Xiao R, Sui X, Song W. Pulmonary sclerosing pneumocytoma with rapid progression: a case description of serial images. Quant Imaging Med Surg 2025;15(11):11679-11684. doi: 10.21037/qims-24-1930