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PULMONARY SCLEROSING PNEUMOCYTOMA: DIAGNOSTIC CHALLENGES, SURGICAL MANAGEMENT AND LONG-TERM OUTCOMES- A NARRATIVE REVIEW WITH AN ILLUSTRATIVE CASE REPORT

Oncology

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ABSTRACT

Introduction : Pulmonary sclerosing pneumocytoma (PSP) is a rare benign epithelial tumor of the lung, accounting for less than 1% of all primary pulmonary neoplasms[12]. It predominantly affects women, particularly of Asian ethnicity, and is most often detected incidentally during evaluation for unrelated conditions[11, 13]. Despite its benign biological behaviour, PSP frequently mimics primary lung carcinoma on radiological imaging, cytology, core needle biopsy, and even frozen section analysis[34]. This overlap creates significant diagnostic challenges and may lead to unnecessarily aggressive surgical resections or inappropriate adjuvant therapies[26]. **Methods :** This narrative review synthesizes available literature, including published studies, representative clinical experience, to provide a comprehensive overview of PSP. Relevant aspects analysed include epidemiology, pathogenesis, clinical presentation, radiological characteristics, histopathology, immunohistochemistry, differential diagnosis, surgical management, and follow-up strategies. **Results :** PSP typically presents as a solitary, well-circumscribed pulmonary nodule, often discovered incidentally. Radiological and cytological features frequently overlap with malignant lung tumors, contributing to diagnostic uncertainty. Histopathological examination, supported by immunohistochemistry, remains the cornerstone for definitive diagnosis. Complete surgical excision is the treatment of choice and is associated with excellent prognosis. Recurrence and metastasis are exceedingly rare, although careful postoperative surveillance is recommended. **Conclusion :** Recognition of PSP and its distinguishing pathological features is essential to avoid misdiagnosis and overtreatment. Accurate diagnosis enables appropriate surgical management, with complete excision being curative in the vast majority of cases. Awareness among clinicians, radiologists, and pathologists is critical to ensure optimal patient outcomes.

KEYWORDS

Pulmonary Sclerosing Pneumocytoma; Sclerosing Hemangioma; Benign Lung Tumor; Pneumocytoma; Lung Carcinoma Mimic.

INTRODUCTION

Pulmonary sclerosing pneumocytoma (PSP), previously referred to as pulmonary sclerosing hemangioma, is an uncommon benign lung tumor first described by Liebow and Hubbell in 1956[7]. At the time of its initial description, the lesion was believed to be of vascular origin due to its prominent hemorrhagic and sclerotic features. However, advances in ultrastructural analysis and immunohistochemistry later demonstrated that PSP originates from primitive respiratory epithelial cells, most likely type II alveolar pneumocytes[1, 3]. This paradigm shift led to its reclassification as an epithelial tumor in the World Health Organization (WHO) classification of thoracic tumors[2].

Despite its benign nature, PSP continues to pose significant diagnostic challenges for clinicians, radiologists, and pathologists. Its radiological appearance often overlaps with malignant pulmonary neoplasms, particularly adenocarcinoma, while its histological heterogeneity may mimic both epithelial and neuroendocrine tumors[34]. Furthermore, rare reports of lymph node metastasis, multifocal pulmonary involvement, pleural dissemination, and delayed local recurrence have raised questions regarding its true biological behaviour[8, 10]. These factors underscore the importance of a thorough understanding of PSP to avoid misdiagnosis and overtreatment.

Epidemiology and Demographic Profile

Pulmonary sclerosing pneumocytoma is rare, accounting for less than 1% of all primary lung tumors[1]. Most published data originate from small case series and retrospective institutional experiences[3, 11]. A striking female predominance has been consistently observed, with reported female-to-male ratios ranging from 4:1 to 5:1[11, 13]. The tumor most commonly presents in middle-aged individuals, typically between the fourth and seventh decades of life[12]. Nevertheless, cases have been reported in younger patients, including those in their second and third decades, and rarely in paediatric populations[14]. Tumor sizes usually range from 1–5 cm, and over 70% of cases are detected incidentally[12].

Geographically, PSP has been reported more frequently in East Asian populations, particularly in China, Korea, and Japan[11, 13]. Whether this represents a true ethnic predisposition or reflects increased diagnostic recognition remains uncertain.

According to the WHO categorization of lung and pleural tumors (2018, ICD-11 for Mortality and Morbidity Statistics), pulmonary sclerosing pneumocytoma was categorized as a benign, fibromatous neoplasm [6]. Nevertheless, several reports have documented possible malignant features, including lymph node metastasis [7–11] or local recurrence [12]. Although the prognosis of pulmonary sclerosing pneumocytoma does not appear to be affected by its malignant potential, other factors may be related to prognosis. Pulmonary

sclerosing pneumocytoma can vary in size and occur in different lobes of the lung[13,14]. However, whether these factors affect prognosis has not yet been examined in a large cohort.

Pathogenesis and Molecular Characteristics

Immunohistochemical studies have been pivotal in elucidating the histogenesis of PSP. Consistent expression of thyroid transcription factor-1 (TTF-1) in both surface cuboidal cells and stromal round cells strongly supports an origin from primitive respiratory epithelium[23-29]. The dual-cell population, a hallmark of PSP, is now considered a manifestation of divergent differentiation from a common epithelial progenitor.

Recent molecular analyses have further supported the neoplastic nature of PSP. Recurrent mutations in the AKT1 gene are the most frequently reported genetic alteration, followed by abnormalities involving the β-catenin signaling pathway[15,16]. These molecular changes are distinct from driver mutations seen in lung adenocarcinoma and likely contribute to the tumor's indolent growth pattern[16].

Clinical Presentation

The majority of patients with PSP are asymptomatic at the time of diagnosis, with lesions discovered incidentally on routine chest radiography or computed tomography performed for unrelated indications[11,12]. When symptoms are present, they are typically mild and nonspecific, including cough, hemoptysis, chest pain, dyspnea, and fatigue[17]. Hemoptysis is thought to result from the tumor's rich vascularity and hemorrhagic components[17].

Rarely, PSP may present as an endobronchial lesion, leading to airway obstruction, recurrent pneumonia, or atelectasis[18]. Multifocal pulmonary involvement has been infrequently reported and may complicate diagnostic interpretation, particularly in patients undergoing evaluation for metastatic malignancy.

Radiological Features

On chest radiography, PSP typically appears as a solitary, well-circumscribed, round or oval pulmonary nodule or mass[20,21].

Computed tomography (CT) usually demonstrates a well-defined lesion with smooth margins and variable internal attenuation. Enhancement patterns may be homogeneous or heterogeneous depending on the proportion of solid, sclerotic, and hemorrhagic components[20,21].

Several radiological signs have been described, including the marginal pseudocapsule sign, overlying vessel sign, air-gap sign, and halo sign[22]. Calcification may be present but is inconsistent, while cavitation is rare.

On FDG-PET imaging, PSP generally shows low to moderate uptake; however, false-positive high uptake has been reported due to hemorrhage, inflammation, and cellularity[23,24], limiting the specificity of PET-CT.

Cytology and Preoperative Diagnostic Limitations

Preoperative diagnosis of PSP remains difficult. Bronchoalveolar lavage is often nondiagnostic. Fine-needle aspiration cytology and core needle biopsy may reveal papillary or glandular epithelial arrangements, leading to misdiagnosis as adenocarcinoma or carcinoid tumor[4,25]. Frozen section analysis is similarly unreliable due to sampling limitations and architectural heterogeneity[26]. Consequently, definitive diagnosis is most often established following complete surgical excision.

As a result, definitive diagnosis of PSP is often established only after complete surgical excision and thorough histopathological evaluation with immunohistochemistry. Awareness of this limitation is crucial to prevent premature therapeutic decisions based on inconclusive preoperative findings.

Histopathology and Immunohistochemistry

Grossly, PSP is a well-circumscribed, firm, unencapsulated tumor with a tan-gray or yellow cut surface. Microscopically, four architectural patterns—papillary, solid, sclerotic, and hemorrhagic—are typically present in varying proportions[27]. A defining feature is the dual-cell population comprising surface cuboidal cells and round stromal cells with bland cytology[28].

Immunohistochemically, both cell types are positive for TTF-1 and epithelial membrane antigen[23,29]. Cytokeratin and Napsin-A expression is limited to surface cuboidal cells, while stromal cells are negative, a feature critical for differential diagnosis.

Differential Diagnosis and Diagnostic Pitfalls

The differential diagnosis of PSP includes both benign and malignant pulmonary lesions. Important considerations include:

1. Pulmonary Adenocarcinoma

Adenocarcinoma represents the most significant diagnostic pitfall due to overlapping glandular and papillary morphology. However, adenocarcinoma lacks the characteristic dual-cell population seen in PSP.

2. Neuroendocrine Tumors

Typical and atypical carcinoid tumors may mimic PSP on limited biopsy specimens. Neuroendocrine marker expression and absence of the classical PSP immunophenotype aid in distinction.

3. Pulmonary Hamartoma

Hamartomas typically demonstrate fat and cartilaginous differentiation radiologically and histologically.

4. Metastatic Lesions

Metastatic papillary carcinomas, particularly from thyroid or renal primary tumors, should be excluded using clinico radiological and immunohistochemical correlation

Surgical Management

Surgical excision remains the cornerstone of management for PSP, serving both diagnostic and therapeutic purposes[5,6]. Limited resections such as wedge resection or enucleation are usually sufficient for peripherally located tumors[32]. Lobectomy may be required for centrally located lesions, large tumors, or when malignancy cannot be excluded intraoperatively.

Although lymph node metastasis has been reported, it does not appear to impact prognosis and should not mandate radical resection[8,33]. Routine completion lobectomy is not indicated once a definitive diagnosis of PSP is established.

CASE REPORT

A 33-year-old female with complaints of low-grade fever for 2 months associated with cough with haemoptysis.

No family history of pulmonary disorders, not a k/c/o DM, HTN, TB
O/E: PS-1/5, RS- B/L normal vesicular breath sounds heard. No palpable chest mass/ lump.

CECT THORAX- large heterogeneous enhancing soft tissue density lesion with surrounding ground glass haziness and septal thickening measuring approximately. 38x38mm seen in the superior aspect of the right lower lobe, (? carcinoma of the lung). Multiple sub centimetric-sized lymph nodes in the pre- and paratracheal areas, most of which are calcified.

BAL fluid cytology- negative for malignancy
Patient was then planned for CT-guided biopsy of the lesion.

Histology s/o glandular epithelial cells lined by cuboidal cells with moderate, clear cytoplasm (pneumocyte-like) and proliferation of round to oval stromal cells.

D/D:
1. Neuroendocrine tumor
2. Sclerosing pneumocytoma
3. Meningioma.

Surgery- RIGHT LOWER LOBE WEDGE RESECTION under GA on 21.01.25

Intra op findings–

- 5x4 cm lesion arising from the right lower lobe upper margin close

to the hilum.
- No pleural effusion.
- Adhesion of lung tissue seen near the biopsy site.

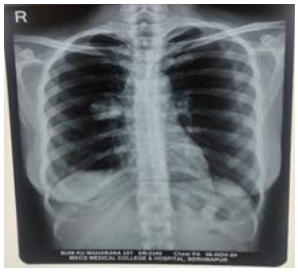


FIG 1: PRE-OPERATIVE

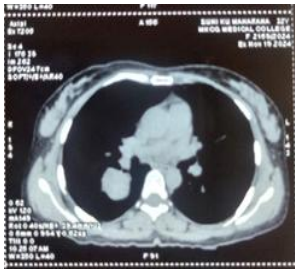


FIG 2: PRE-OPERATIVE CECT THORAX

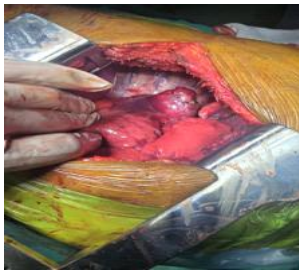


FIG 3: INTRA OP

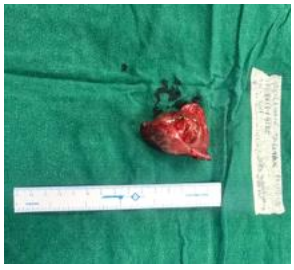


FIG 4: FINAL SPECIMEN

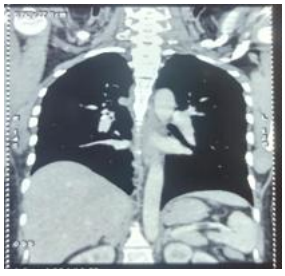
Histopathology: Papillary solid and haemorrhagic pattern composed of cuboidal surface cells and round stromal cells with foamy and hemosiderin-laden macrophages in stroma suggestive of sclerosing pneumocytoma. Round cells surrounded by surface epithelial cells in a papillary pattern, Hypercellular areas with foamy macrophages. Pan CK negative in central round cells, positive in surface epithelial cells, TTF 1 nuclear positive in both peripheral epithelial cells and central round cells

Follow-up CECT thorax done after 3 months s/o no fresh lesion. Patient is clinically doing well.

Follow-up x-ray after 6 months s/o no fresh lesion. Patient is clinically doing well.

Prognosis, Metastasis, and Recurrence

PSP carries an excellent prognosis following complete surgical excision, with survival rates comparable to those of the general population[5,11]. Rare cases of lymph node involvement, multifocal disease, pleural dissemination, and delayed recurrence have been reported[8–10,34]. Recurrence is exceedingly uncommon and is usually attributed to incomplete resection rather than aggressive tumor biology[9,10,34].



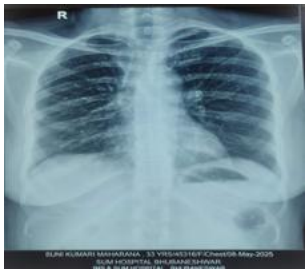


FIG 5: POST- OP CECT THORAX AFTER 3 MONTHS
FIG 6: POST- OP X-RAY AFTER 6 MONTHS

Postoperative Follow-Up and Surveillance

Given rare reports of delayed recurrence, postoperative surveillance is advisable. Most authors recommend imaging at 6 months postoperatively, followed by annual chest imaging for at least 3–5 years[6,10,34]. Extended surveillance may be considered in cases of multifocal disease or when margins are uncertain.

CONCLUSION

Pulmonary sclerosing pneumocytoma is a rare benign epithelial lung tumor that frequently mimics malignancy, leading to diagnostic uncertainty and potential overtreatment. Accurate diagnosis relies on awareness of its characteristic histopathological and immunohistochemical features. Complete surgical excision is curative in nearly all cases and is associated with excellent long-term outcomes. Increased recognition of this unique pulmonary neoplasm will help prevent misdiagnosis and unnecessary aggressive therapy.

Consent

Informed written consent was obtained from the parents for the present case report

Ethical Approval

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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