

Large Symptomatic Sclerosing Pneumocytoma in a Young Male Smoker—A Rare and Deceptive Presentation



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Pulmonary sclerosing pneumocytoma (PSP) is a rare pulmonary tumor that behaves in a benign fashion and has excellent prognosis.¹ The tumor was historically termed as sclerosing hemangioma by Liebow and Hubbell in 1956.² The incidence peaks in sixth decade with a striking predilection for females (F:M = 5:1).^{3,4} Although it presents as a well-circumscribed solid nodular mass, the radiological features are nonspecific and can be misconstrued as malignant especially in large tumors.⁵ Frozen sections and trucut biopsy interpretation of such tumors may also be misleading.

A 26-year-old male, with a short-term smoking history, presented initially to other institute with complaints of chest heaviness and mild pain for the last 6 weeks. Contrast-enhanced computed tomography thorax revealed a well-defined solid pulmonary mass lesion in right upper lobe measuring 4.3 × 3 × 2 cm. Few subcentimetric mediastinal lymph nodes were noted (→Fig. 1). Excision was done through right posterolateral thoracotomy, which was reported as non-small cell carcinoma-adenocarcinoma (NSCC-ADC); pathological stage pT2N cannot be assessed.

The patient then subsequently presented to our institute for further management with radiological and histopathological review. Hematoxylin and eosin sections revealed a well-circumscribed unencapsulated nodular tumor with a mixture of growth patterns: papillary, solid, sclerotic, and

hemorrhagic (→Fig. 2A–D). The tumor had two cell types—cuboidal surface cells and oval-to-round stromal cells (→Fig. 3A–B). The complex papillae had cuboidal surface cells, while oval-to-round stromal cells were in a streaming pattern. Few surface cells also showed clear vacuolated cytoplasm, multinucleation, and intranuclear inclusions. Also evident were foci of mature fat (→Fig. 2C), calcification and areas with hemosiderin pigment, foamy macrophages, and cholesterol clefts. There was no evidence of high-grade cytological atypia or nuclear pleomorphism. Mitosis was sparse.

On immunohistochemistry (IHC), the surface epithelial cells were diffusely positive for pancytokeratin (CK) and thyroid transcription factor 1 (TTF1), whereas the underneath stromal cells were positive for only TTF1 (→Fig. 3D) and negative for cytokeratin (CK) (→Fig. 3C). Paired-box gene 8 (PAX8) was absent in both the components. Overall, clinical, radiological, and histopathological features provided tangible evidence for the final diagnosis of PSP. The patient is on 12 months of follow-up with no evidence of disease.

PSP is characteristically a small tumor, typically asymptomatic, detected incidentally and shows a wide age range (11–80 years) distribution, commonly seen in middle-aged Asian women.^{1,3,4} The cell of origin for this tumor has remained enigmatic and has changed over time and was previously conceived as vascular neoplasm.² Now, it is seemingly derived

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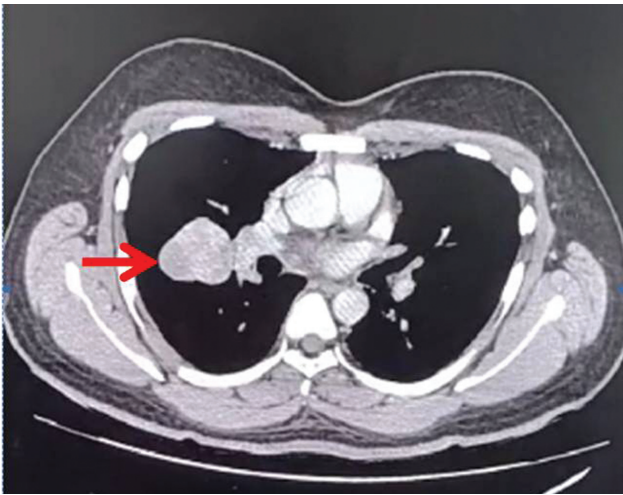


Fig. 1 Contrast-enhanced computed tomography thorax showed relatively well-defined nodular lobulated mass lesion (→) in right upper lobe.

from primitive pulmonary epithelium. As per the molecular studies, a monoclonal pattern has been demonstrated in round and surface cells. AKT1 internal tandem duplications, point mutations, and short indels have been recognized in nearly all

sclerosing pneumocytomas.^{1,5} Shin et al⁵ have reviewed clinico-radiological characteristics of 76 cases of PSP. The median age was 50 years, and 88.2% of patients were females. Only 7.9% patients were symptomatic for the mass and had hemoptysis, while in 71% it was incidentally detected and in 21.1% it was detected during workup for metastasis for other malignancy. Great majority of the patients had single lesion (92.1%) and the mean diameter was 2.27cm. The cases with size more than 3cm, with irregular borders, were more prone for being misdiagnosed as lung cancer. Rarely, lymph node metastases can also happen.⁵

Four histological patterns can be seen in typical PSP: papillary, sclerotic, solid, and hemorrhagic.³ The papillary and solid histological pattern can be misdiagnosed as papillary NSCC-ADC and carcinoid, especially on frozen section or small diagnostic biopsy.¹ Judicious IHC panel is helpful for resolving the differential diagnosis of papillary ADC and carcinoids of lung (See [Table 1](#) for differential diagnosis). TTF1 is expressed in both surface epithelial and stromal cells of PSP and it is also positive in papillary ADC and carcinoid tumors of lung, hence can be misleading. In PSP, cuboidal surface and round stromal cells are both epithelial membrane protein (EMA) and TTF1 positive. Pancytokeratin, CK7, Napsin A stain surface cells and the round stromal cells are

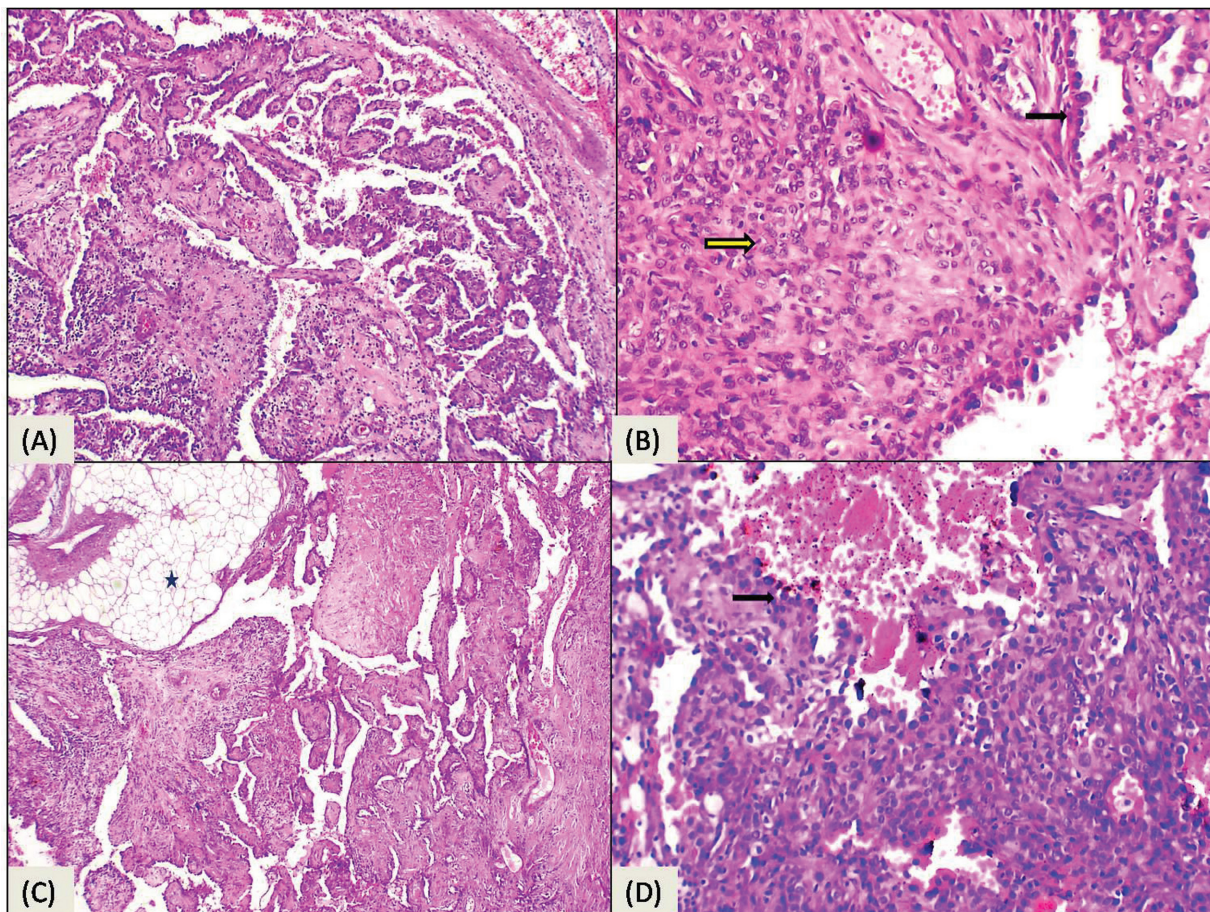


Fig. 2 Hematoxylin and eosin stain shows mixture of growth patterns in sclerosing pneumocytoma. (A) Papillary architecture that mimics true papillae. (B) Solid architecture with sheets of round stromal cells (⇒) with tubule (⇨) lined by surface cells. (C) Sclerotic papillae, fibrosis with hyalinized collagen admixed with a component of mature fat (★). (D) Hemorrhagic pattern with blood filled spaces lined by cuboidal epithelium (⇨).

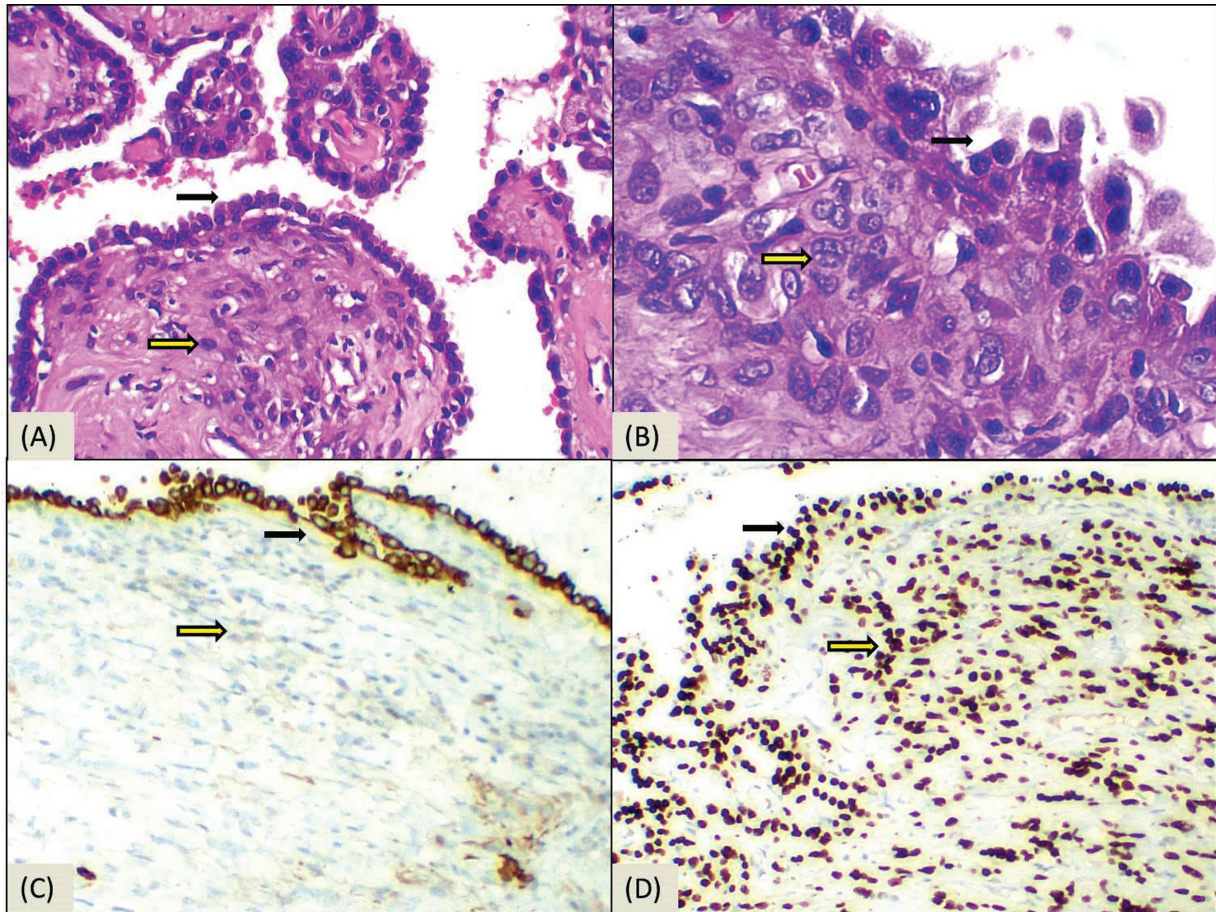


Fig. 3 Sclerosing pneumocytoma. Images (A) and (B) show two tumor cell types: surface cuboidal cells (➡) lining the papillae and round-to-oval stromal cells (➡). (C) Strong cytokeratin expression in surface cuboidal cells (➡) while negative in round-to-oval stromal cells (➡). (D) Strong thyroid transcription factor 1 expression in both surface cuboidal cells (➡) and round-to-oval stromal cells (➡).

either negative or weakly positive. Differential and contrast immunostaining of surface epithelial cells and negative-to-weak positive immunorexpression in stromal cells by CK are very helpful for confirmation.

Detailed clinical history, diligent histomorphological, and IHC examination espoused with correlation of clinical and radiological findings like absence of significant lymphadenopathy as evaluated in the presented case are imperative as

Table 1 Radiological and pathological features of different entities

Entity	Radiological	Histological	IHC
Pulmonary sclerosing pneumocytoma	No distinct radiological features have been described in literature; however, some CT signs may be useful such as the marginal pseudocapsule sign, the most common sign	Well-circumscribed tumor with varied growth patterns: papillary, solid, sclerotic, and hemorrhagic. The tumor had two cell types—cuboidal surface cells and oval-to-round stromal cells. The cuboidal surface cells are similar to type II pneumocytes and the round cells are small with central bland nuclei. No significant cytologic atypia or nuclear pleomorphism seen. Mitosis is rare	Surface and round stromal cells are both epithelial membrane protein (EMA) and TTF1 positive. Pancytokeratin, CK7, Napsin A stain surface cells but the round stromal cells are negative or weakly positive
Papillary adenocarcinoma of lung	No distinct radiological features have been described in literature	The tumor consists of neoplastic glandular cells growing along the surface of fibrovascular cores. The neoplastic cells display significant cytologic atypia, nuclear pleomorphism and frequent mitosis	Tumor cells are CK, TTF1 and Napsin A positive

Table 1 (Continued)

Entity	Radiological	Histological	IHC
Carcinoid tumor of lung	Central tumors are usually seen as a single hilar or perihilar mass that is usually well-defined, often marked homogeneous contrast enhancement due to high vascularity; calcification (usually eccentric) can occur but is not a common feature	Various organoid growth patterns with trabecular, rosette formation, palisading, ribbon, or solid arrangements. Papillary and true glandular patterns can be seen Tumor cells are monotonous uniform, fine granular chromatin, inconspicuous nucleoli Graded according to mitosis and necrosis	Tumor cells are CK, synaptophysin, chromogranin, CD56, and INSM1 positive TTF1 tends to be positive in peripheral carcinoids but negative in central tumors

Abbreviations: CT, computed tomography; IHC, immunohistochemistry; TTF1, thyroid transcription factor 1.

PSP is treated by surgical resection alone that is considered as curative.^{1,3,4}

Informed Consent

Informed consent was obtained from all individual participants included in the study.

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Conflicts of Interest

None disclosed.

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