

Mini Review

A Rare Pulmonary Tumor Mimicking Lung Cancer:
Sclerosing PneumocytomaElktaibi A^{1*}, Pirel M¹, Marty-Ané C², Serre I¹ and Costes Martineau V¹¹Department of Pathology, Gui de Chauliac Teaching Hospital, France²Department of Cardiac, Thoracic Vascular Surgery, Arnaud De Villeneuve Hospital, France***Corresponding author:** Elktaibi A Department of Pathological Anatomy Gui de Chauliac Teaching Hospital, France**Received:** August 07, 2019; **Accepted:** August 26, 2019; **Published:** September 02, 2019**Abstract**

Pulmonary sclerosing pneumocytoma is an uncommon benign tumor of the lung with a predilection for middle-aged females. Diagnosis is often made incidentally following chest computed tomography scan performed for systematic check-up. We report a case of Pulmonary sclerosing pneumocytoma in a 53 year-old female who got admitted for medical control of her malignancy. Radiology a single, well-circumscribed mass infiltrating the right lung. Histological examination showed a tumor which was composed of mixture patterns with solid, cystic, and papillary structures. A panel of immunohistochemical markers was performed and the tumor was finally diagnosed to be sclerosing pneumocytoma.

Keywords: Sclerosing pneumocytoma; Histopathology; Surgery**Introduction**

Pulmonary Sclerosing Pneumocytoma (PSP) is a rare benign lung tumor, which was previously reported and named sclerosing hemangioma in 1956 by Liebow [1]. It was categorized as a 'miscellaneous tumor' according to the 1999 and 2004 World Health Organization classification of lung tumors [2]. In 2015, the most recent WHO classification changed the name of this tumor. It is now called sclerosing pneumocytoma and is classified in the category of adenomas. Due to its rarity, the clinical symptoms and preoperative imaging performance lacks specificity [3]. In the present study, we report a case of PSP in patient with history of malignancy due to its rare occurrence and to investigate the clinicopathological characteristics with a brief review of the literature

Case Presentation

A 53-year-old woman with remarkable medical history: - Osteosarcoma of the left femur diagnosed and treated in 2006 - Papillary carcinoma of the thyroid treated in 2012 and followed by mammary carcinoma in 2017 with good therapeutic response. She was presented for a medical checkup. The computed tomography scan of the thorax demonstrated a 26 mm nodular isolated lesion in the lower lobe of the right lung (Figure 1). Clinically, a metastatic lesion was suggested because of the patient's history. To confirm the diagnosis, wedge resection of the right lower lobe was performed and sent to pathological department for extemporaneous analysis. Gross examination demonstrated a well-circumscribed lesion, solid and cystic mass with hemorrhagic areas (Figure 2). The frozen section diagnosis was mesenchymal proliferation of undetermined malignant potential. After formalin fixation, microscopic observation showed three morphological patterns. A papillary pattern was predominant, composed of cuboidal surface cells with benign features. The solid pattern of growth comprising sheets of round stromal cells with occasional epithelial/surface cells. A cystic component with hemorrhage was also noticed (Figure 3). Immunohistochemically, surface cuboidal cells are positive for Cytokeratin 7 (CK7), where as stromal round cells are focally positive for CK7. Both



Figure 1: Contrast-enhanced chest tomography reveals a well-defined mass in the right lower lobe (arrow).

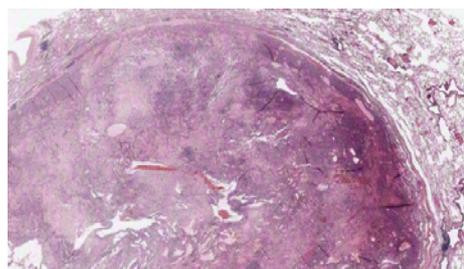


Figure 2: Low-power view of the well-circumscribed tumor with surrounding normal lung parenchyma (hematoxylin-eosin, ×4 magnification).

populations express Thyroid Transcription Factor-1 (TTF-1) and Epithelial Membrane Antigen (EMA). Progesterone showed nuclear immunoreactivity in the round stromal cells (Figure 4). This case was assigned to the molecular biology platform but no mutation was found in NGS (*Next Generation Sequencing Panel INCa V2*) technique. At the fourth month post-surgery follow-up appointment, the patient was alive with no evidence of recurrence or metastasis.

Discussion

PSP is a rare neoplasm of the lung first described by Liebow and Hubbell in 1956 [1]. It has a marked female preponderance (male to

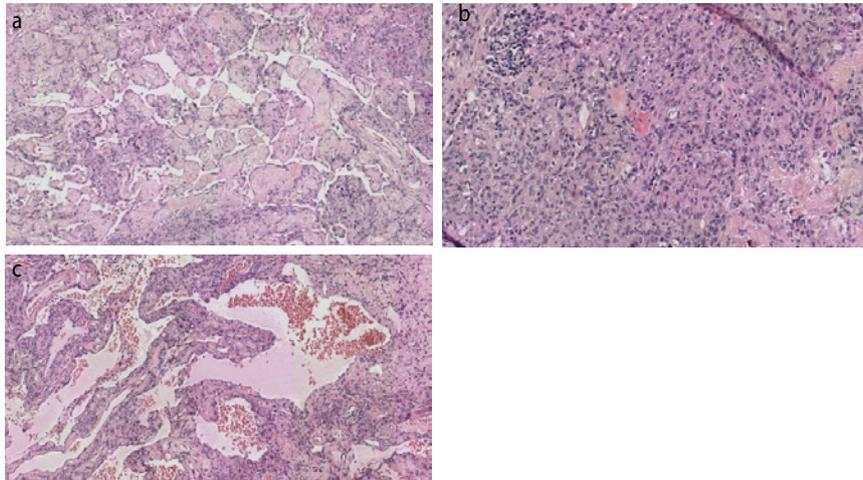


Figure 3: Histological patterns of sclerosing pneumocytoma; (a)- Papillary pattern formed by cuboidal surface cells and round stromal cells with sclerotic core; (b)- Solid pattern; (c)-Cystic and hemorrhagic pattern (hematoxylin and eosin , x 40).

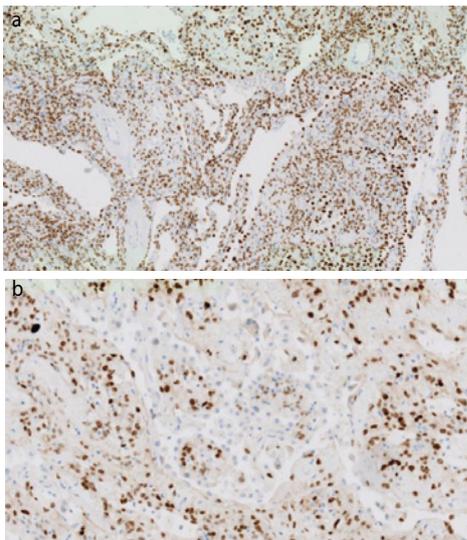


Figure 4: An immunohistochemical analysis showing positive expression of (a)- TTF-1 in both surface and round cells and (b)- nuclear immunoreactivity of progesterone in the round stromal cells with no reactivity of the surface cuboidal cells (hematoxylin and eosin , x 40).

female ratio 1:5) [4], and its incidence appears to be higher in East Asia than in western countries. The tumor can occur in all age groups, and the average is 46 years [5]. The tumor size ranges from 0.3 to 8 cm and 73% of lesions are smaller than 3 cm [6]. Clinically, most patients are asymptomatic and the disease is usually found incidentally on a radiograph [4]. Radiologically, the tail sign was described as a key CT finding of pulmonary sclerosing pneumocytomas. It was defined by a small linear projection seen radiating out from the lesion [3]. Histologically; four architectural patterns have been described: papillary, solid, sclerotic, and hemorrhagic. Typically, the tumor is composed of the combination of the four patterns and is constituted by superficial cuboidal and round interstitial cells. Pulmonary sclerosing pneumocytoma can be very challenging to diagnose in frozen sections, small biopsies, and cytology where they can easily be mistaken for adenocarcinoma, carcinoid tumors [3], or metastasis

especially in patients with malignancy history. In the present case, the correct diagnosis was made on the surgical resection specimen after paraffin inclusion. These tumors typically have a benign clinical course, with excellent prognosis and no recurrence. A few cases of pneumocytoma lymph node metastasis have been reported; however, this result did not affect the prognosis [7]. Surgical resection is recommended for sclerosing pneumocytoma; nevertheless, dilemma exists about lobectomy or sublobar resection. Some data reported similar recurrence-free survival after lobectomy and sublobar resection [8]. For other authors, PSP presents a benign behavior, and a simple enucleation is sufficient [5].

Conclusion

Sclerosing pneumocytoma is a rare lung tumor that is usually recognized as a solitary nodule in the lung. CT scanning can suggest its benignity; although radiological features are not specific for a definite differential diagnosis from other primary or metastasis pulmonary tumors. Surgical removal is recommended; however an accurate preoperative diagnosis of sclerosing pneumocytoma based on the analysis of specimen obtained by small biopsies proves very helpful for a benign tumor whose limited surgical excision is curative.

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