



Huge Pulmonary Sclerosing Pneumocytoma with Endobronchial Invasion: A Case Report with a Literature Review

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ARTICLE INFO

Received February 16, 2021

Revised April 6, 2021

Accepted April 30, 2021

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Pulmonary sclerosing pneumocytoma (PSP) is a tumor of pneumocytic origin that is classified as a benign neoplasm. To date, aggressive behavior of this tumor has rarely been reported. Here, we describe a case of a 56-year-old woman with a huge, 19-cm PSP that resulted in mediastinal shift and showed microscopic endobronchial invasion and necrosis. The differential diagnosis included malignant mesenchymal tumors, such as solitary fibrous tumor; however, PSP was confirmed based on the characteristic thyroid transcription factor 1 positivity and membranous expression of Ki-67 on immunohistochemical staining of tumor cells.

Keywords: Pulmonary sclerosing hemangioma, Computed tomography, Immunohistochemistry, Case report

Case report

Pulmonary sclerosing pneumocytoma (PSP), also known as pulmonary sclerosing hemangioma, is a rare benign pulmonary neoplasm that predominantly affects middle-aged adults, with a significant predilection for women [1]. PSP was formerly thought to have a vascular origin; however, it was later revealed that it originates from the epithelium. Thus, it was reclassified as an adenoma by the World Health Organization. The size of PSP ranges from 0.3 to 7.0 cm, and 73.7% of the lesions are smaller than 3 cm [2]. On rare occasions, huge tumors are seen, and it is difficult to recognize the accurate epicenter, leading to an incorrect differential diagnosis. Herein, we report a case of a huge PSP presenting with mediastinal shift, endobronchial invasion, and necrosis.

This case report was approved by the Institutional Review Board of Gangnam Severance Hospital (3-2020-0445). The requirement for informed consent was waived due to the retrospective nature of this study.

A 56-year-old woman presented with dyspnea and cough. Chest radiography showed an abnormal shadow

over the entire left side of the chest and mediastinal deviation to the right (Fig. 1A). The initial contrast-enhanced chest computed tomography (CT) revealed a mass, about 19 cm in size, that almost filled the entire left thoracic cavity, causing a rightward mediastinal shift. The mass showed heterogeneous enhancement and a central low-density area. Due to its large size, the tumor protruded into the left main bronchus, almost reaching the level of the carina (Fig. 1B). This endobronchial lesion was also confirmed by bronchoscopy (Fig. 1C). On positron emission tomography, the huge, well-defined mass showed diffusely increased glucose metabolism, central defects, and strong fluorodeoxyglucose uptake (Fig. 1D). However, an accurate diagnosis was not possible because only a small amount of degenerative tissue was seen on transbronchial biopsy, and re-biopsy was not possible due to the patient's condition. Surgical treatment was selected considering the patient's severe respiratory symptoms. Left pneumonectomy was performed through median sternotomy. This method was chosen (instead of left thoracotomy) to secure a surgical view and easily approach the hilar structure, as well as to allow the use of cardiopulmonary bypass or ex-



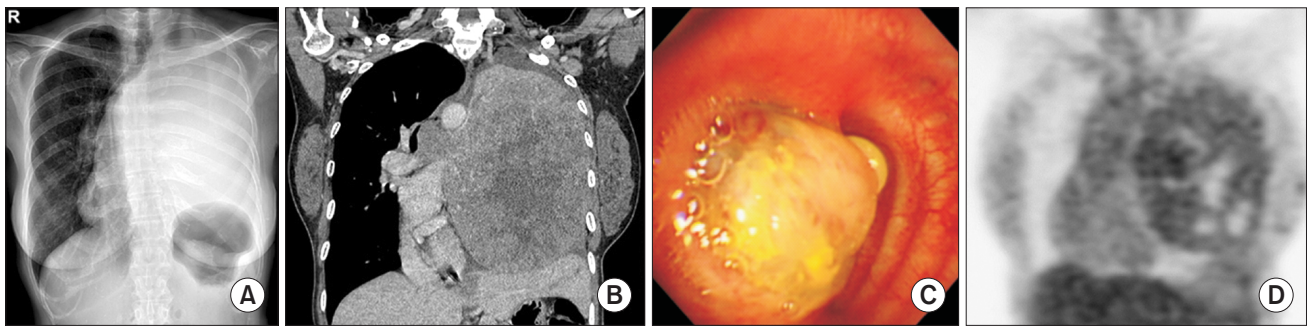


Fig. 1. (A) Posteroanterior-view chest radiograph shows a huge mass in the left hemithorax with complete atelectasis of the left lung and rightward deviation of the mediastinum. (B) Chest computed tomography image with coronal reformation reveals a low-density area in the center of the tumor, suggesting hemorrhage and necrosis. Endobronchial extension of the tumor is also noticed. (C) Endobronchial lesion in the left main bronchus confirmed by bronchoscopy. (D) Positron emission tomography image shows a well-defined, huge mass with diffusely increased glucose metabolism, central defects, and strong fluorodeoxyglucose uptake.

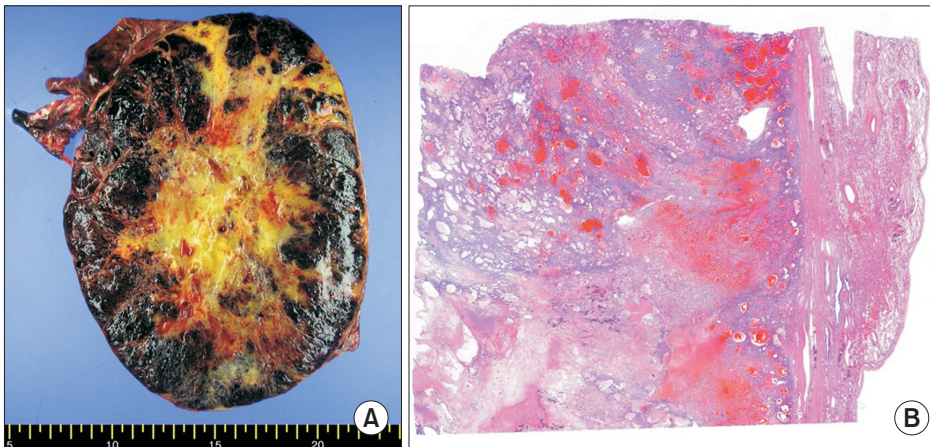


Fig. 2. (A) Cut surface of the mass shows a well-circumscribed, hemorrhagic, tan mass with central degenerative changes. (B) Scan power view shows extensive hemorrhagic lakes with cellular and acellular areas. Necrosis is seen in the left lower part.

tracorporeal membranous oxygenation in the event of possible bleeding or unstable vital signs during surgery. Grossly, the entire left lung was replaced by the tumor mass, with adhesions throughout the mediastinal space and pleural cavity. The mass almost extended up to the carina, and thus, we had to transect the proximal part of the left main bronchus. There was a residual mass on the left main bronchus. No attempts were made to remove it since PSP is a benign lesion, and we postulated that it would eventually undergo necrosis and self-expectoration, as it had no blood supply.

On gross examination, a huge, relatively well-defined mass, measuring 19.5×13.0×12.0 cm, replaced the entire left lung. The cut surface of the mass was diffusely hemorrhagic and yellowish, and hyalinized degenerative changes were observed in the central area (Fig. 2A).

On microscopic examination, an overlap between the cellular and acellular areas, multifocal hemorrhagic and cystic changes, and necrosis of the degenerative area were

observed (Fig. 2B). The tumor cells showed a mixture of papillary and sclerotic patterns (Fig. 3A). On immunohistochemical staining for thyroid transcription factor 1 (TTF-1), signal transducer and activator of transcription 6 (STAT6), and Ki-67, the tumor cells were diffusely and strongly positive for TTF-1 and lacked STAT6 expression (Fig. 3B). Additionally, a membranous staining pattern was observed on manually performed Ki-67 staining (Fig. 3C). The tumor labeling index of Ki-67 was 1%–2%. In the area near the bronchus, tumor cells infiltrated into the bronchial subepithelium (Fig. 3D). Additional immunohistochemical staining for cytokeratin (CK) delineated epithelial cells of the acinar pattern, while stromal cells lacked CK expression (Fig. 4A, B). Epithelial membrane antigen was positive in both the epithelial and stromal components (Fig. 4C). No tumor was found in the bronchial resection margin; however, protruding and floating tumor cells were found within the bronchial lumen. Based on the results of histological testing and immunohistochemical staining, a final

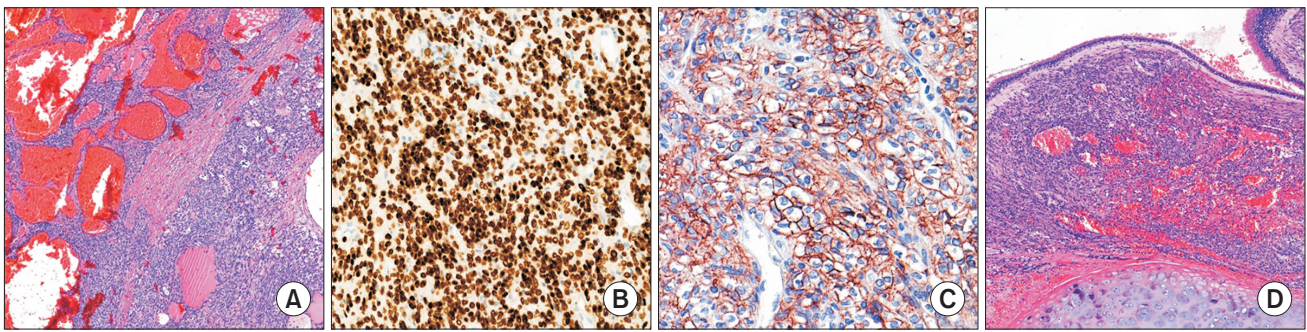


Fig. 3. (A) Solid pattern and sclerosis associated with a blood lake (left). (B) Immunohistochemical staining of tumor cells for thyroid transcription factor 1 is diffusely and strongly positive. (C) Membranous expression of Ki-67 on manual staining, which is a characteristic feature of sclerosing pneumocytoma. (D) Tumor cells invading the bronchial subepithelium.

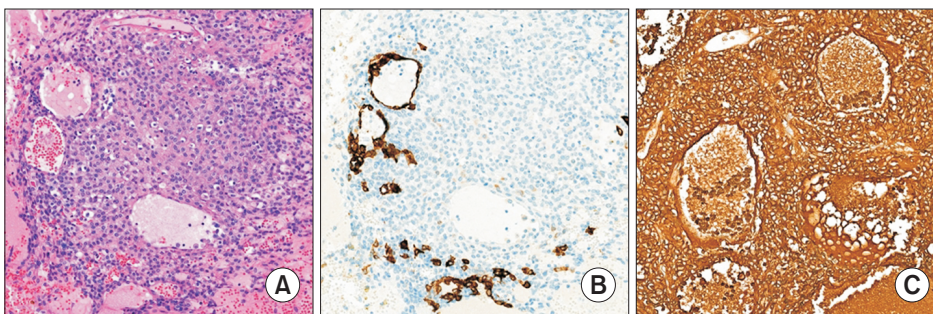


Fig. 4. (A) Epithelial component of an acinar pattern with an intervening stromal cell component. (B) Epithelial component shows strong positivity for cytokeratin immunohistochemistry. (C) Both epithelial and stromal components are positive for epithelial membrane antigen immunohistochemistry.

diagnosis of PSP was made. Postoperatively, the patient was on mechanical ventilation in the intensive care unit. She was extubated on the first postoperative day and discharged on the seventh postoperative day.

Discussion

PSP, formerly known as pulmonary sclerosing hemangioma, is a rare benign pulmonary neoplasm that predominantly affects middle-aged adults. It was formerly thought to be a variant of hemangioma that originates from the mesenchymal cells [3]; however, it is now considered an epithelial tumor composed of a dual population (surface cells and round cells) [2,4].

PSP appears to have a marked female predominance (male-to-female ratio of 1:5). While most patients with PSP are asymptomatic, some may present with dyspnea, cough, chest pain, or hemoptysis, depending on tumor location and size. Although PSP is categorized as a benign neoplasm, a few cases showing malignant features, such as local recurrence and lymph node metastasis, have been reported. Devouassoux-Shisheboran et al. [2] reported that hilar lymph node metastasis occurred in approximately 1% of patients with PSP.

On CT, PSP typically presents as an inhomogeneous enhancing soft tissue mass with a smooth outline ranging from 67–112 Hounsfield units [5]. These imaging features correlate well with the histological characteristics of PSP. High-density, isodense, and low-density areas represent hemorrhagic, solid/sclerotic, and cystic tumor components, respectively [5]. Mixed areas of high and low signal intensity are seen on T1- and T2-weighted magnetic resonance images, and intense enhancement is seen on enhanced T1-weighted magnetic resonance images.

Most commonly, the size of PSP ranges from 0.3 to 7.0 cm; 73.7% of them are smaller than 3 cm [2]. However, there have been reports of PSPs larger than 3 cm, making the diagnosis challenging [6]. This case, in line with the latter, exhibited uncommon findings; the size of the tumor in our case was extraordinarily large (19.5 cm), which made it difficult to identify its origin. Large PSPs with sizes of 15 cm, 11 cm, and 10 cm have been reported [6]. To the best of our knowledge, the tumor in our case was larger than any PSP reported to date. Furthermore, we noted an endobronchial lesion. These features are rarely associated with PSP, further complicating the differential diagnosis.

The name “pulmonary sclerosing hemangioma” was derived from its sclerosing, angiomatoid, and papillary fea-

tures, but it is now considered to originate from pneumocytes. Most patients with PSP have a benign clinical course; however, in fewer than 20 cases, lymph node metastasis has been reported [7]. Patients with lymph node metastasis tended to have large tumors that were located in the left lung [7]. These factors may increase the malignant potential of PSP.

Complete surgical excision with clear margins is commonly accepted as the standard treatment for PSP, and there is no evidence supporting the effectiveness of neo-adjuvant or adjuvant therapies. It has been reported that a surgically unresectable PSP with encasement of the pulmonary artery was successfully treated with radical external-beam radiation therapy [8]. PSP is generally associated with a benign clinical course and shows an excellent prognosis; no tumor-related mortality has been reported in the literature.

In summary, we report a rare case of PSP with clinically aggressive features, such as a huge size and endobronchial invasion.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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