

Pulmonary Sclerosing Pneumocytoma with Lymphnode Metastasis: A Case Report

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ABSTRACT

Pulmonary sclerosing pneumocytoma is a rare tumor originated from primitive respiratory epithelial cells. It's a relatively benign tumor of the lung but has potential malignancy according to literatures reported. Herein, we reported a 42-year-old man who received video-assisted left upper lobectomy in our hospital on 22nd December 2016. The post operation pathology suggested pulmonary sclerosing pneumocytoma with peribronchial lymph nodes metastasis. The patient recovered well from operation.

Keywords: Lymphnode metastasis, pulmonary sclerosing pneumocytoma, rare tumour, malignancy

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INTRODUCTION

Pulmonary sclerosing pneumocytoma (PSP, formerly named as pulmonary sclerosing hemangioma) is a rare tumor of the lung first described by Leibow and Hubbell in 1956 (1). It was once considered to derive from endothelial cells while now accepted as originating from primitive respiratory epithelial cells. The tumor mainly comprises of two types of cells, lining cells and round cells. Lining cells were epithelial cells with multiple morphologies, resembling type II alveolar cells. Round cells distributed in the interstitium, with oval or polygonal in shape (2). PSP mostly occurred in middle-aged female in Asia. Female-to-male ratio reached to 5:1 as reported by Devouassoux–Shisheboran after studying 100 cases of PSP (3). It was generally considered as a benign tumor, but several reports manifested that PSP has potential of malignancy. Tumor cells can metastasize to mediastinal lymph node (4) and even bone (5). However, PSP with lymph node metastasis are very rare all over the world. Herein, we reported a 42-year-old man who received video-assisted left upper lobectomy in our hospital and the diagnosis was pulmonary sclerosing pneumocytoma with peribronchial lymph nodes metastasis after surgery.

CASE REPORT

A 42-year-old man came to our hospital with chest distress, cough and faint yellow sputum for one month, without fever or chest pain. Chest computed tomography (CT) scan showed a high-density oval mass in the left upper lobe. The mass was lobulated and the internal density was nonuniform (Figure1). Positron emission computed tomography (PET-CT) showed an irregular

mass in the left upper lobe with increased metabolic rate of fluorodeoxyglucose. The standard uptake value reached to 3.5. Fiberoptic bronchoscopy was normal and no malignant cell histology was found. Tumor markers such as Alpha-fetoprotein (AFP), Carcinoembryonic antigen (CEA), Carbohydrate antigen 199 (CA199), squamous cell carcinoma antigen (SCC-Ag), neuron-specific enolase (NSE), Cytokeratin-19-fragment were all in normal range. The patient had a smoking history of 20 cigarettes per day for 20 years.

He was diagnosed as pulmonary carcinoma and received video-assisted left upper lobectomy in our hospital on 22nd December 2016. Intraoperative frozen examination indicated pulmonary sclerosing pneumocytoma. Then mediastinal lymph node dissection was performed. The patient recovered well and discharged 4 days after surgery.

Microscopic examination showed tumor cells diffusely distributed in the mass, with interstitial blood vessels dilated and congested. The tumor cells stained positively for thyroid transcription factor-1 (TTF-1), epithelial membrane antigen (EMA), cytokeratin 7 (CK7). Tumor cells were detected in two of the eight peribronchial lymph nodes near hilum (Figure2-3). The final pathological diagnosis after operation was pulmonary sclerosing pneumocytoma with lymph nodes metastasis.

DISCUSSION

Pulmonary sclerosing pneumocytoma was a rare pulmonary tumor once considered to derive from endothelial cells. Due to the development of diagnostic technology such as immunohistochemistry and electron microscopy, the disease is now widely accepted as an

epithelial neoplasm. The tumor mainly contains two type of cells: lining cells and round cells. TTF-1 and EMA were found positive in both cells (2, 6), which not only indicated the epithelial origination of the tumor but also the diagnostic value of the markers. The tumor was formerly named as pulmonary sclerosing hemangioma. The term “sclerosing hemangioma” was changed to “sclerosing pneumocytoma” and the tumor was moved from miscellaneous tumors to the adenoma category in the 2015 World Health Organization Classification (7).

The disease mostly occurred in middle-aged female. The symptoms varied from cough, sputum, chest pain, hemoptysis, repeated bloody sputum to asymptomatic. Devouassoux-Shisheboran et al retrospectively studied 100 cases of PSP. The female-to-male ratio was reported to be 5:1. The tumor located in the right lung in 52 patients (54%), compared with 44 patients (46%) in the left lung. One case was found with peribronchial lymph nodes metastasis (3). YASUSHI ADACHI et al found there were 18 patients with lymph node metastasis reported all over the world (8). The data were collected and compared to that of 100 cases reported by Devouassoux-Shisheboran et al. The mean age of metastasis group was 36 years, younger than the 46 years of the 100 cases. Female to male ratio was 9:8 of the 18 cases. Mean tumor size of the metastasis group was 44.8mm compared with 26mm of the 100 cases. The result indicated that PSP with lymph node metastasis seemed to occur more often in patients of young age or with larger tumor size. The reason may be these tumors grow more rapidly.

Surgical resection should be performed once PSP was diagnosed or suspected. The outcome is often excellent even if there is lymph node metastasis. Joon Seok Park et al retrospectively reviewed 32 patients who received limited resection or lobectomy for PSP. They found no difference in patients characteristics and surgical results between the two groups (9).

However, this is a small research and further study should be performed with larger cohort. Due to the malignant potential, mediastinal lymph node should be dissected. PSP was reported to be sensitive to radiation therapy. Russell WB Fayers reported a 45-year old man with unresectable PSP of the left lung. The patient underwent definitive radical external beam radiation therapy and achieved almost complete metabolic tumor response, no evidence of metastasis was found (10).

In conclusion, pulmonary sclerosing pneumocytoma is a rare tumor with relatively benign nature but has potential malignancy. Mediastinal lymph node metastasis can be detected in very rare cases. Surgical resection is effective and excellent outcome can be achieved after surgery. Lymph node dissection should be performed during operation due to the potential malignancy of pulmonary sclerosing pneumocytoma.

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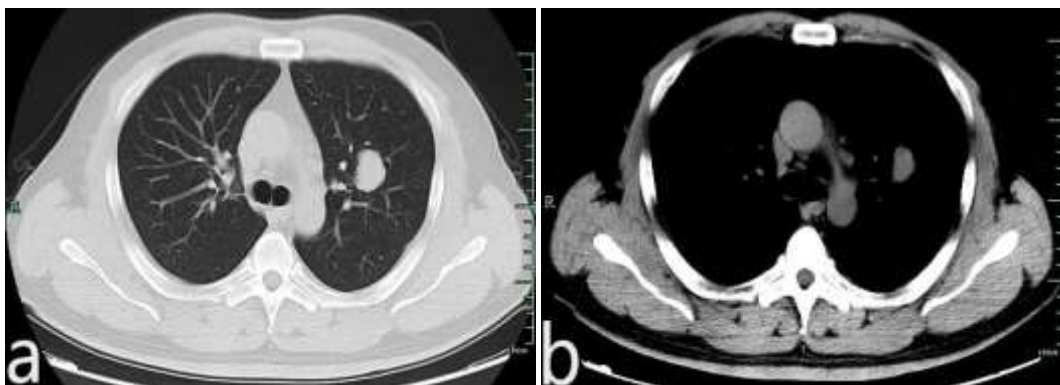


Fig. 1: Chest CT scan of the patient.

CT scan showed a high-density oval mass in the left upper lobe. The mass was lobulated and the internal density was nonuniform. (a) pulmonary window; (b) mediastinal window.

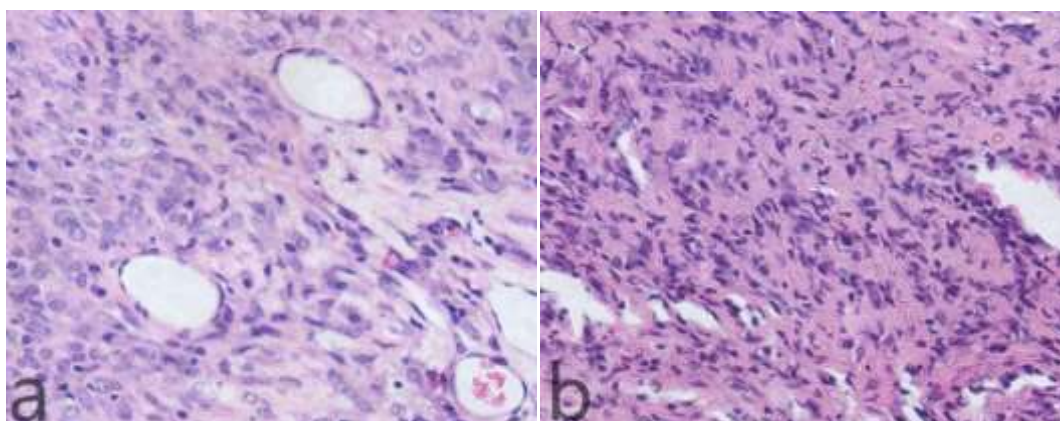


Fig. 2: Hematoxylin-eosin (HE) staining of the tumor and metastatic lymph node.

Hematoxylin-eosin (HE) staining of the tumor and metastatic lymph node.

Pulmonary sclerosing pneumocytoma in left upper lobe (HE×20)

Pulmonary sclerosing pneumocytoma metastasized to lymph node. (HE×20)

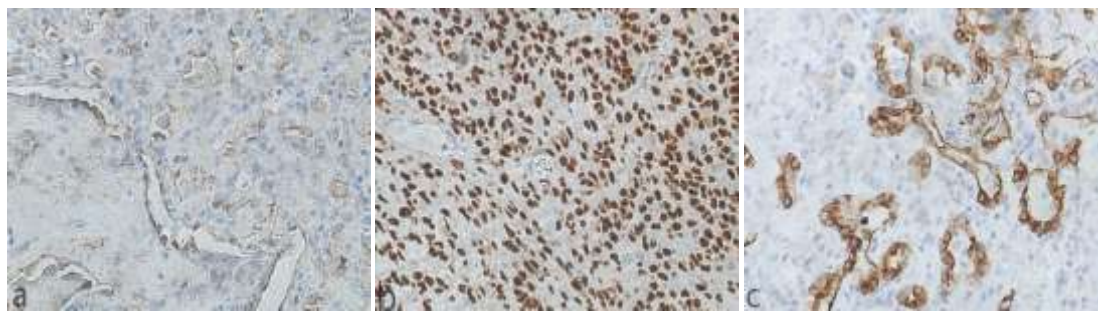


Fig. 3: Immunohistochemistry staining of the tumor.

Immunohistochemistry staining of the tumor. Immunohistological staining for PSP in left upper lobe was positive with (a) epithelial membrane antigen, (b) thyroid transcription factor-1, (c) cytokeratin 7 (magnification, $\times 20$).