Bilateral Multiple Pulmonary Sclerosing Pneumocytomain A Young Male: Case Report

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Case report

Keywords: Pulmonary nodule, Pulmonary sclerosing pneumocytoma, Lung adenocarcinoma, Lung surgery, Multiple

Posted Date: August 4th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-686381/v1

Abstract

Background: Pulmonary sclerosing pneumocytoma (PSP) is a relatively rare benign tumor, and multiple pulmonary sclerosing pneumocytomas are even rarer. PSP is usually seen in adults over 50 years old, featuring a female to male ratio of 5:1. We report an extremely rare case of a 20-year-old male patient with bilateral multiple pulmonary sclerosing pneumocytoma. This rare benign disease also presents diagnostic challenges throughout the treatment process.

Case presentation: A 20-year-old male was diagnosed with bilateral multiple pulmonary nodules on chest CT. The patient underwent regular chest CT reexamination over the course of six months, which indicated a tendency of slow enlargement of pulmonary nodules. After complete examination and exclusion of tumors in other parts of the body, he underwent two separate surgeries, and bilateral multiple nodules were removed. Intraoperative frozen section analysis suggested that some nodules were adenocarcinoma and the remaining nodules were considered benign tumors. Postoperative immunohistochemistry indicated that all nodules were pulmonary sclerosing pneumocytomas.

Conclusions: Bilateral multiple pulmonary sclerosing pneumocytoma are rare in young men. In clinical practice, the differentiation of this disease from malignancy is challenging, and surgical treatment is a safe option for pulmonary nodules with a tendency to become larger.

Background

Pulmonary sclerosing pneumocytoma (PSP) is a rare benign tumor. Clinically, PSP of the lung is most frequently encountered in middle-aged women, with a peak incidence at 50 years of age(1). Typically, PSP presents with a single nodule, and multiple nodules are very rare. In addition, the diagnosis of sclerosing alveolar cell tumor remains challenging, both radiologically and histologically. We report an extremely rare case of a 20-year-old male patient with bilateral multiple pulmonary sclerosing pneumocytoma. This case also presents diagnostic challenges throughout the treatment process.

Case Presentation

The patient, a 20-year-old male, was admitted to the hospital due to multiple nodules in both lungs. Prior to hospitalization, the patient underwent regular chest CT reexamination over the course of six months, which indicated a tendency of slow enlargement of pulmonary nodules. The patient's physical examination showed no obvious abnormalities, and there was no previous family history of tumor. No significant increase in tumor markers was observed in blood samples. Enhanced chest CT showed multiple high-density nodules in both lungs with clear boundaries (Figs. 1, 2). The largest nodule was located in the right lower lobe, measuring approximately 2.8 cm * 2.3 cm. Enhanced chest CT showed obvious homogeneous enhancement of the nodules. No enlarged lymph nodes were found in the mediastinum or hilum. PET-CT showed multiple nodules in both lungs, increased uptake of the imaging agent, and SUVmax: 4.8-5, no obvious metabolic abnormality was found in other parts of the body

(Fig. 3). A fine-needle aspiration (FNA) cytology of the right lower pulmonary nodule revealed a large number of epithelioid cells with small cell atypia, and malignancy could not be ruled out.

Considering the young age of the patient, the tendency of pulmonary nodules to become larger, and the fact that the current examination could not rule out malignant tumor, we decided to remove the pulmonary nodules surgically. We performed thoracoscopic right middle lung lobectomy + right upper lung wedge resection + right lower lung wedge resection under general anesthesia(Fig. 4). intraoperative frozen sections showed that there were microinvasive adenocarcinomas in the right upper lung (0.4 cm in diameter), two nodules in the right middle lung (both 0.9 cm in diameter), and two nodules in the right lower lung (2.1 cm in diameter). Postoperative immunohistochemistry indicated that all nodules were pulmonary sclerosing pneumocytomas(Fig. 5), and the hilar and mediastinal lymph node samples were negative. The patient recovered smoothly and was discharged from the hospital. One month later, he returned to the hospital for left lung surgery. He underwent thoracoscopic wedge resection of the left upper lung and left lower lung. Postoperative immunohistochemistry showed all nodules were pulmonary sclerosing pneumocytomas: CK-Pan (+), EMA (+), TTF-1 (+), VIM (+), P40 (-). The patients recovered and was discharged smoothly.

Discussion

Pulmonary sclerosing pneumocytoma (PSP), formerly known as pulmonary sclerosing hemangioma, is a rare pulmonary tumor initially described by Liebow et al(2). in 1956, as a tumor with marked sclerosis and vascularization. PSP is usually seen in female over 50 years old, and most of them are single nodules. In the 2015, the World Health Organization (WHO) classification “miscellaneous tumors” was changed to “adenomas”(3). The essential feature of PSP is the presence of cuboidal surface cells and round stromal cells, which consist of four major histological patterns; hemangiomatous, papillary, sclerotic, and solid(3). The immunohistochemical staining pattern is positive for TTF-1 and EMA(4). Our case is a 20-year-old young man with bilateral multiple PSP, which has rarely been reported.

Over the last years, many studies have discussed the origin, differential diagnosis, potential malignancy and best treatment option for PSP. For many years, in fact, this tumor was presumed to be of vascular origin. Instead, immunohistochemical results, in particular the TTF-1 expression, confirm its epithelial origin from the respiratory epithelium(4). Because of the unspecific radiological features mimicking malignancies and its histological heterogeneity, the differential diagnosis with adenocarcinoma and carcinoid tumors is still challenging. Patients are usually asymptomatic and it is detected incidentally. Chest CT scan usually reveals a round, solitary, well-circumscribed, homogeneous nodule and FDG PET/CT scan usually reveals a low to moderate uptake in PSPs(5). Our patient also had no clinical symptoms, and chest CT and PET-CT findings were similar to those reported in the literature. From the imaging inferences, the pulmonary nodules of the patient were considered benign first. We also performed fine-needle aspiration (FNA) cytology on the largest nodules, but no evidence of malignant tumor was found. Therefore, we followed the patient for 6 months, and found that the pulmonary nodules had a tendency to slowly grow larger. In view of this situation, we decided to perform surgical treatment

for the patient. Pre-operative diagnosis, when feasible, is helpful to plan the best therapeutic strategy. Gal et al. was the first to report that the cytologic diagnosis of PSP requires the identification of its dual cell population, made up of abundant stromal cells and fewer surface cells(6). The problem is that fine-needle aspiration (FNA) cytology is poorly diagnostic, both due to the rarely distinct separation of the two tumor cell types in the specimen and to the disease rarity and hence potential unfamiliarity with its cytologic features of most pathologists. Preoperative cytologic and histological findings of PSP are limited to few case reports, in which computed tomography (CT)-guided FNA, EBUS-TBNA or intraoperative frozen sections (FS) often led to misdiagnosis(7). In our case, intraoperative frozen section suggested PSP coexisting with adenocarcinoma, but immunohistochemical results revealed that all nodules were sclerosing alveolar cell tumor. PSP combined with lung adenocarcinoma has indeed been reported in very few reports(8, 9). PSP can also be combined and admixed with other lung tumors, such as mucinous adenomatous hyperplasia(10), glandular papilloma (GP)(11), pulmonary spindle cell carcinoma (PSCC)(12), extensive neuroendocrine lesions (including pulmonary neuroendocrine cell hyperplasia, multiple carcinoid tumorlets and typical carcinoid tumors within one pulmonary lobe)(13). FNA cytology is poorly diagnostic, and the misdiagnosis rate of intraoperative frozen section is also high. Shang Z et al. reports the rate of intraoperative misdiagnosis of PSP smaller than 1 cm was 11.1%(14). The pathological diagnosis of PSP is challenging, and obtaining sufficient samples and immunohistochemical examination is the key to improve the diagnosis rate.

PSP can be associated with other tumors at different sites, such as hereditary nonpolyposis colorectal cancer (HNPCC, Lynch syndrome)(15), familial adenomatous polyposis(16), retroperitoneal liposarcoma and renal carcinoma(17), multiple uterine fibromas, cysts of the thyroid and kidney(18), and endometrial stromal sarcoma(19). This indicates that other malignant tumors can coexist with multiple solid pulmonary nodules and be misdiagnosed at times as multiple pulmonary metastases, which would affect the clinical treatment decision; therefore, these masses need to be carefully identified. It has been reported that PSP may present with hilar and mediastinal lymph node metastasis and pleural dissemination(20–22). In the present case, the hilar and mediastinal lymph nodes were not significantly enlarged, and the pathological results from the hilar lymph nodes were negative.

Even with the help of enhanced CT, PET-CT, puncture biopsy or intraoperative rapid pathological reports, PSP may be misdiagnosed. Surgical resection is the treatment of choice for PSP because there have been no reports of recurrence in the literature. Lymph node dissection should be considered for huge tumors because lymph node metastases correlate with a larger tumor size(23). The chest CT of this patient revealed 7 nodules, 3 in the left lobe and 4 right. We resected all the nodules through two surgeries, and the patient recovered smoothly each time. We will perform a long follow-up to assess recurrence and metastasis for this patient.

Conclusions

Bilateral multiple PSPs are very rare in young men. The clinical diagnosis of sclerosing alveolar cell tumor is challenging, and a knowledge of this rare type of neoplasm is fundamental, for both surgeons and

pathologists. Surgery is an important method for diagnosis and treatment of sclerosing alveolar cell tumor.

List Of Abbreviations

Pulmonary sclerosing pneumocytoma (PSP)

glandular papilloma (GP)

pulmonary spindle cell carcinoma (PSCC)

hereditary nonpolyposis colorectal cancer (HNPCC, Lynch syndrome)

Declarations

Ethics approval and consent to participate This study has been approval and consented by the patient. This study was approved by the Medical Ethics Committee of Jinhua Central Hospital.

Consent for publication The work described was original research that has not been published previously, and not under consideration for publication elsewhere, in whole or in part.

All the authors listed have approved the manuscript for publication and copyright transfer of this paper.

Written informed consent for publication was obtained from all participants.

Availability of data and materials Not applicable

Competing interests The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding The present study was supported by the Jinhua Science and Technology Bureau (2020-3-041-1).The funder subsidized the English language editing servicesand publishing expenses of this paper.

Author ContributionsLXS was responsible for patient management and was a major contributor in writing the manuscript. HSQ contributed to data collection, data analysis and bibliography retrieval. CXG contributed to case conception, revising for important intellectual content and final approval ofreport, supervising surgeon. All authors read and approved the final manuscript.

Acknowledgements Not applicable

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Figures

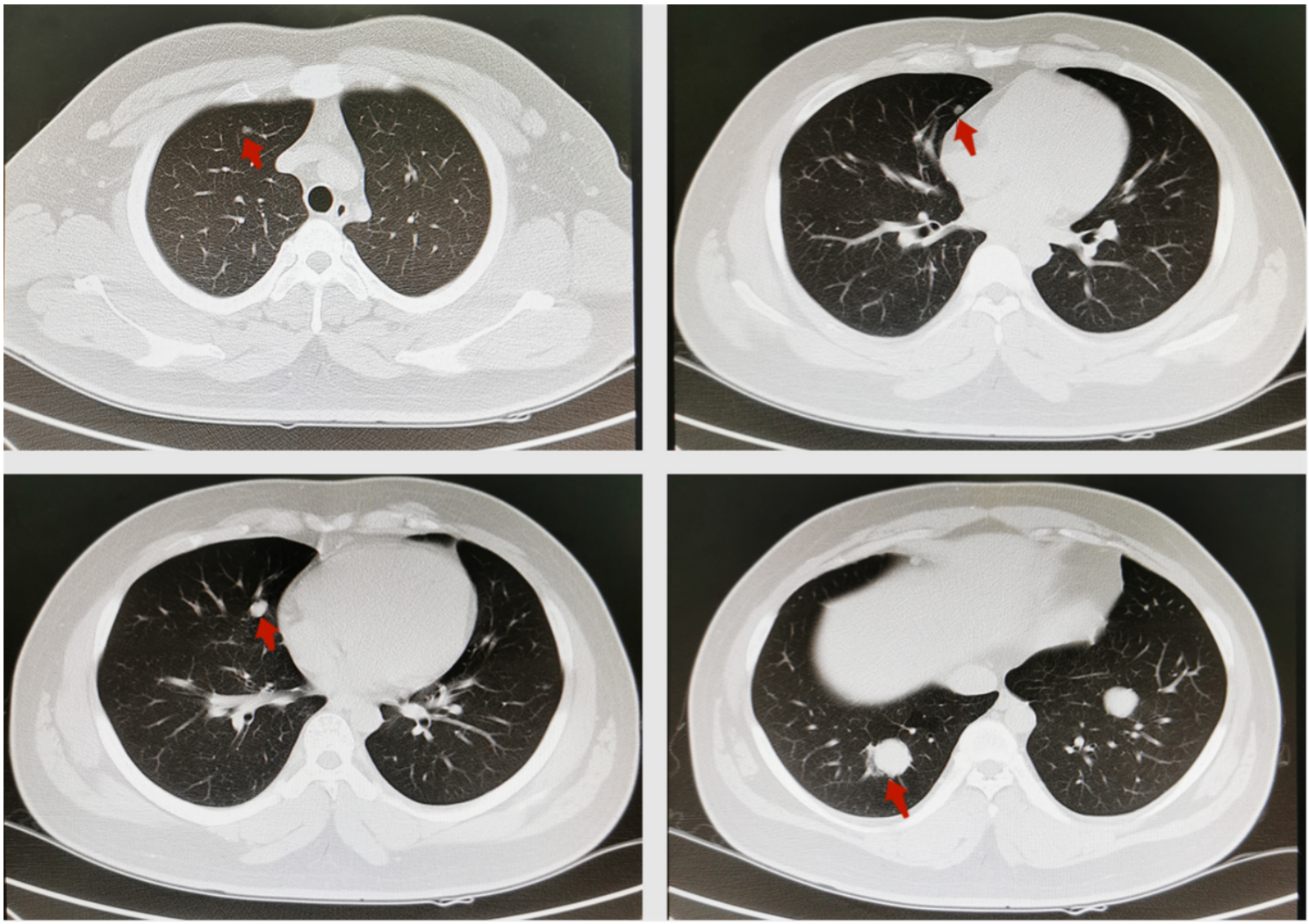

Figure 1

Location of left pulmonary nodules in the first operation(The arrows point to the nodules)

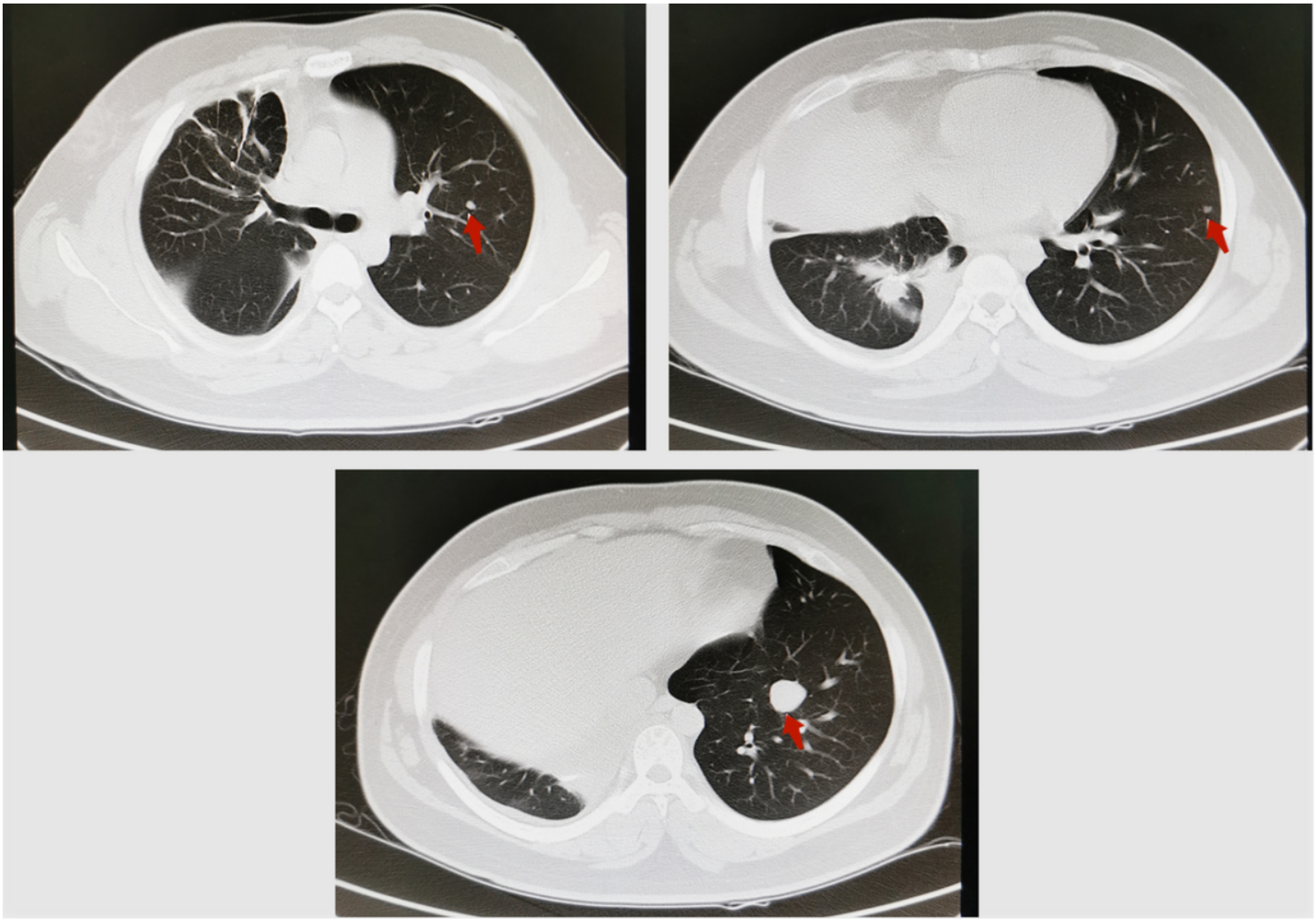

Figure 2

Location of right pulmonary nodules in the second operation(The arrows point to the nodules)

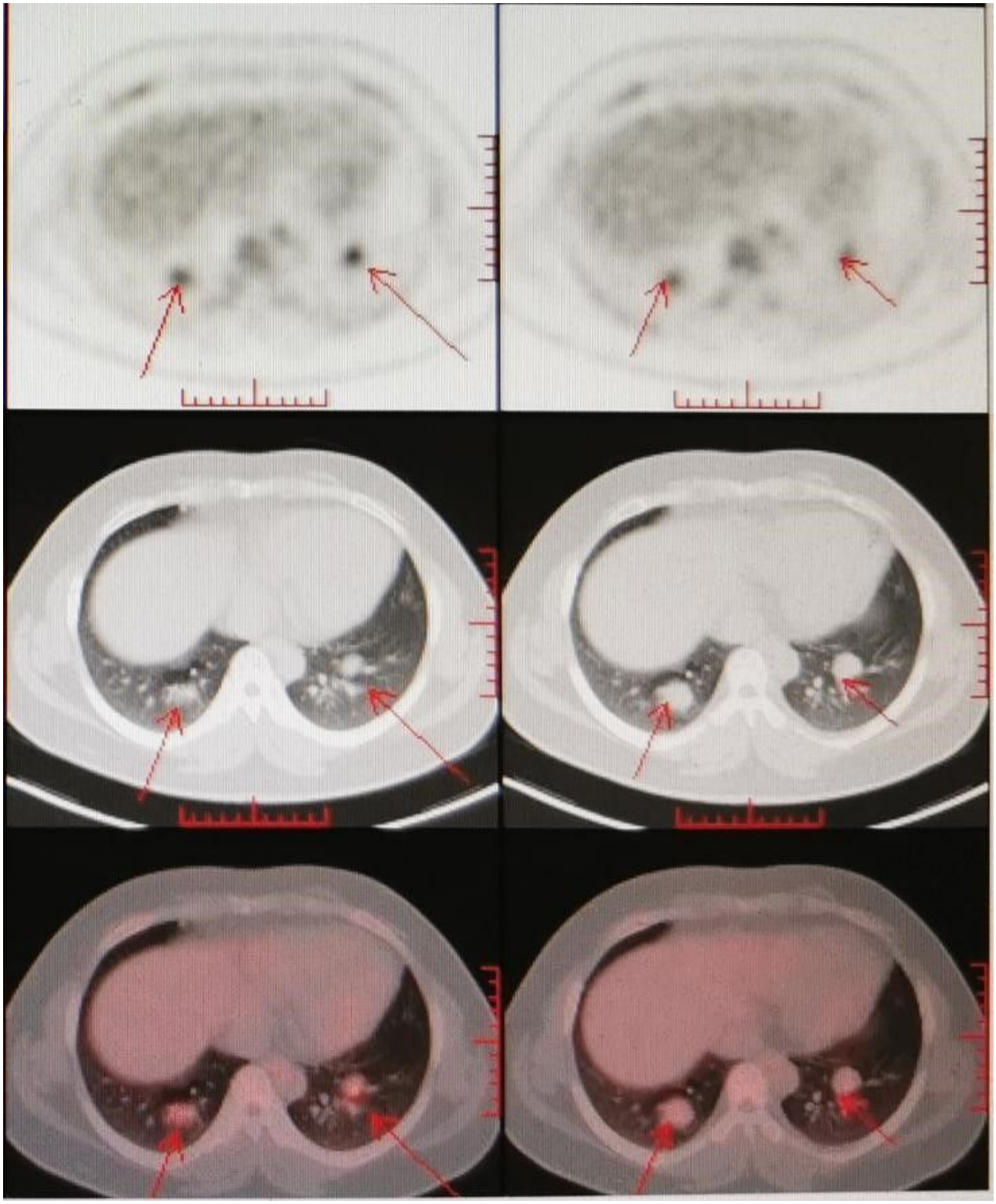

Figure 3

PET-CT images show multiple nodules in both lungs, increased uptake of imaging agent, and SUVmax: 4.8-5(The arrows point to the nodules)

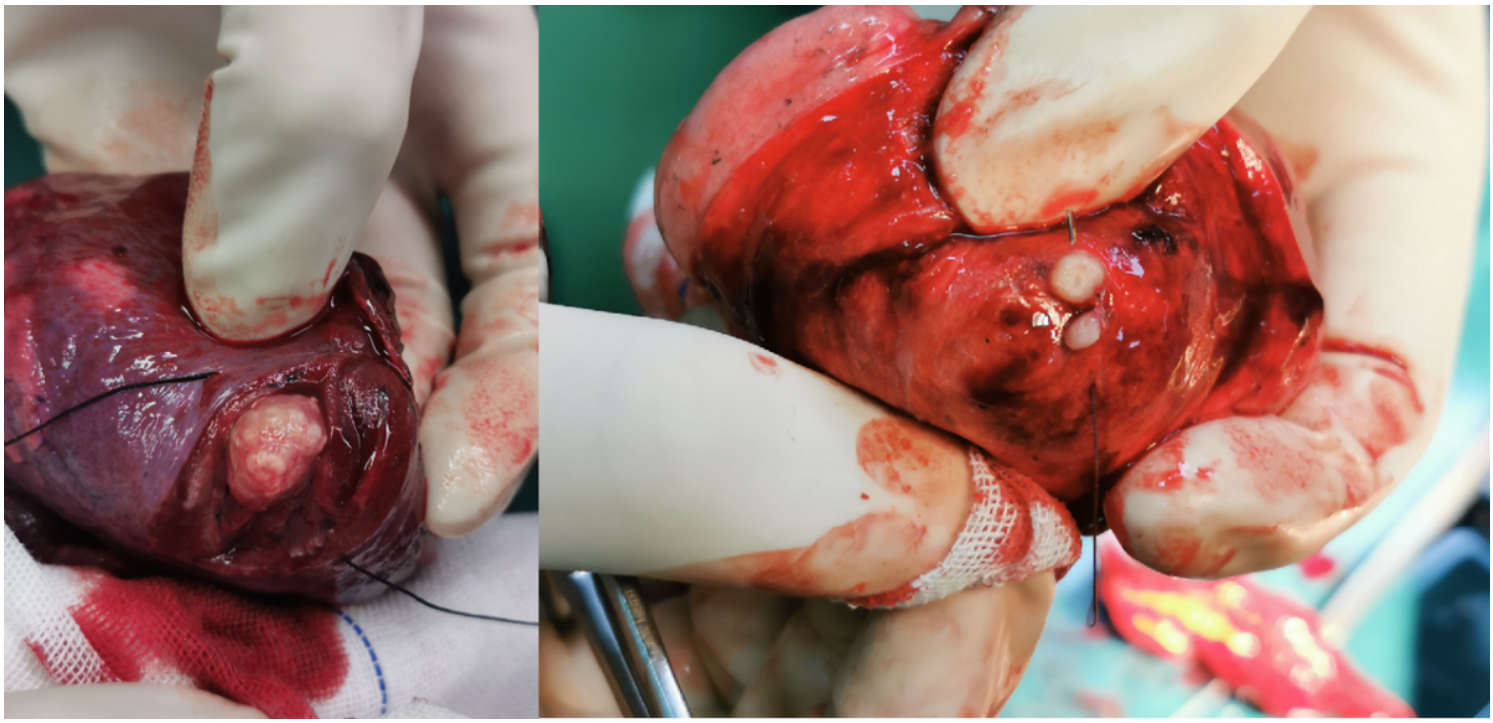

Figure 4

Photographs of surgical specimens of PSP

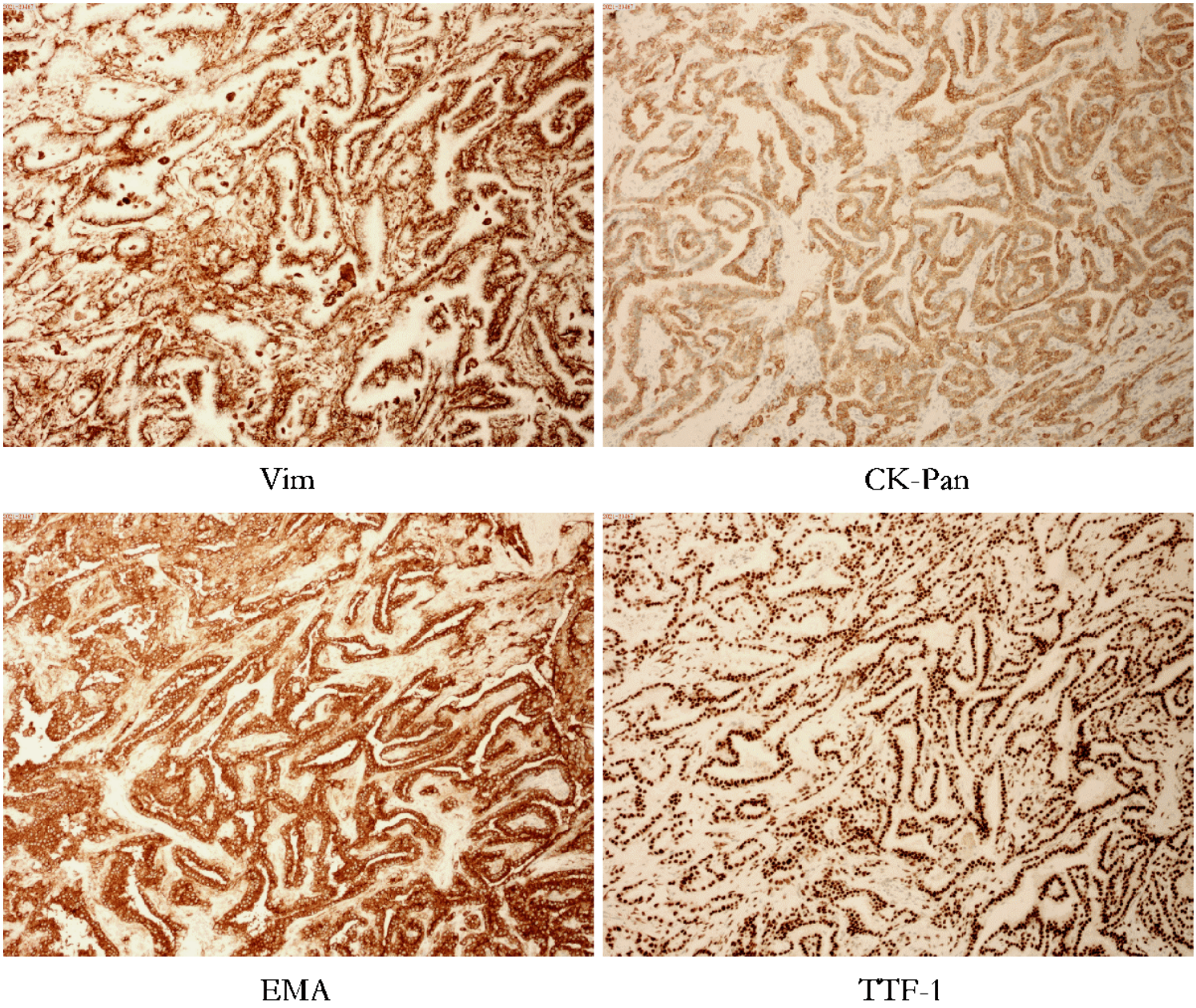

Figure 5

Postoperative immunohistochemistry of PSP,CK-Pan (+), EMA (+), TTF-1 (+), VIM (+)