Maric et al., J Clin Case Rep 2015, 5:10
DOI: 10.4172/2165-7920.1000608

Journal of Clinical Case Reports

Case Report Open Access

# Pulmonary Sclerosing Haemangioma - Case Report

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## Abstract

**Introduction:** Pulmonary sclerosing haemangioma (PSH) is a benign tumour, also known under the term "pneumocytoma". It is a rare disease, of low grade of malignancy possibly originating from type II pneumocytes or Clara cells. It is mostly notified in older females and is usually solitary. According to the histological appearance, sclerosing haemangioma could be: papillary, sclerotic, solid and haemorrhagic.

**Case Report:** A 40-year old female patient, smoker, was hospitalised because of the surgical curing mostly asymptomatic lesion in the lower right lobe. The lesion has been radiologically diagnosed a few years before, and regularly controlled by a Computed Tomography (CT). During the last control, the growth of the lesion was noted and the surgical curing indicated. After clinical examinations, by using Video Assisted Thoracoscopy (VATS), the exploration and precised identification of the well vascularised lesion was performed and the atypic resection of the whole tumour was done. The tumour was hystologically established as sclerosing pneumocytoma, containing two types of cells cuboidas and ovoid, with the same immunophenotype. The following controls have pointed out neider recidive nor malignant alternation.

**Conclusion:** Sclerosing haemangioma should be considered in patients with radiologically diagnosed "coin–like" lesions of a solid as well as multiple type. The complete surgical resection is considered the only effective treatment for PSH, and the normal pulmonary tissue should be reserved as possible.

**Keywords:** Pneumocytoma; Haemangioma; Coin lesion; Pulmonary neoplasm

## Background

Pulmonary sclerosing hemangioma (PSH) is a rare benign tumor of the lungs [1]. PSH frequently occurs in the asymptomatic middle-aged women, predominantly in Asian women in their fifth and sixth decades of life [2].

Devouassoux-Shisheboran et al. [3] analyzed 100 cases of PSH, including one with lymph node metastasis. Patients ranged in age between 13 and 76 years (mean, 46 years). There were 83 female and 17 male patients; thus, the female-to-male ratio was 5:1 [3].

Various scholars have suggested that this tumor originates from vascular endothelial, mesothelial, mesenchymal, epithelial, or neuroendocrine cells. Recently, other reports have provided evidence supporting the theory that PSH originates from type II pneumocytes.

Although PSH is categorized as a miscellaneous tumor according to the 2004 World Health Organization classification of lung tumors, its etiology and origin remain controversial [4].

PSH is considered to be evolved from primitive undifferentiated respiratory epithelium [2].

These tumors are composed of cuboidal surface cells and polygonal stromal cells and show four histological manifestations: hemorrhagic, papillary, solid, and sclerotic. Most tumors show at least three of these histological characteristics [4].

## Case Report

A 40-year old female patient, smoker, was hospitalised because of the surgical curing mostly asymptomatic lesion in the lower right lobe. The lesion has been radiologically diagnosed a few years before, and regularly controlled by a Computed Tomography (CT). During the last control, the growth of the lesion was noted and the surgical curing indicated. Chest X-ray figured out a coin like lesion of up to 15 mm in maximal diameter in the lower right lobe (Figure 1). Chest CT scan showed a well circumscribed node in the posterior segment of the lower right lobe, without mediastinal lymphadenopathy (Figures 2 and 3).

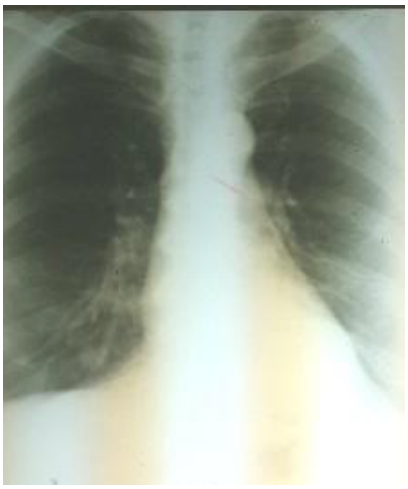

**Figure 1:** Chest X-ray figured out a coin like lesion of up to 15mm in maximal diameter in the lower right lobe.

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**Received** September 01, 2015; **Accepted** October 17, 2015; **Published** October 24, 2015

**Citation:** Maric N, Kostovski V, Pandrc MS, Cvijanovic V, Ristanovic A, et al. (2015) Pulmonary Sclerosing Haemangioma - Case Report. J Clin Case Rep 5: 608. doi:10.4172/2165-7920.1000608

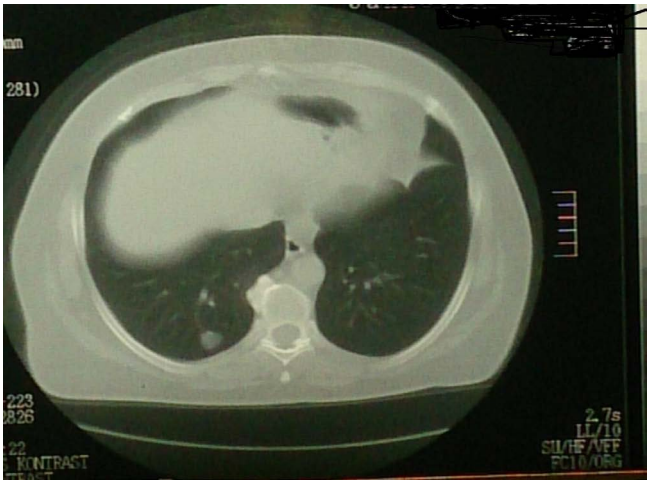

**Figure 2:** Chest CT scan showing a well circumscribed node in the posterior segment of the lower right lobe, without mediastinal lymphadenopathy.

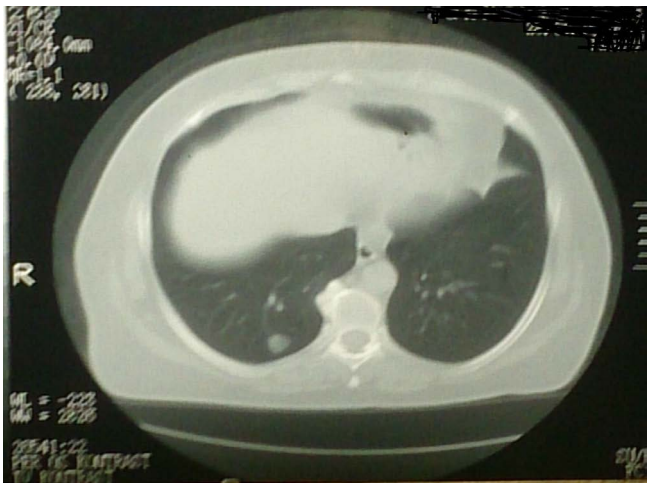

**Figure 3:** Chest CT scan showing a well circumscribed node in the posterior segment of the lower right lobe, without mediastinal lymphadenopathy.

After clinical examinations, by using Video Assisted Thoracoscopy (VATS), the exploration and precised identification of the well vascularised lesion was performed and the atypic resection of the whole tumour was done. The tumour was solid, sclerotic, well vascularised, of up to 11 mm in maximal diameter.

The tumour was hystologically established as sclerosing pneumocytoma, containing two types of cells cuboidas and ovoid, with the same immunophenotype. Initially, Periodic acid Schiff and Periodic acid Schiff stains were done in order to exclude alveolar soft part sarcoma and sugar like pulmonary tumour. By using immunophenotypisation, the presence of the antibodies to the following antigens was documented: Epithelial Membrane Antigen (EMA), Thyroid Nuclear Factor 1 (TTF1), cytokeratins (pan CK), surfactant A, that confirmed the diagnosis.

The following controls have pointed out neither recidive nor malignant alternation.

## Discussion

PSH can occur in any individual in the age group of 4 to 70 years [5,6]. PSH is predominant in 50 year olds, with a female to male ratio of 5:1 in this patient group [7]. Most of the PSH patients are asymptomatic, and only some show clinical manifestations, including cough, hemoptysis, chest pain, and stuffiness. The tumor often occurs at the intralobar site, and less commonly in the bronchus and mediastinum [4]. Devouassoux-Shisheboran et al. analyzed 100 cases of SH, including one with lymph node metastasis. The left lung was the site of 46% of tumors (17% in the left upper lobe, 25% in the left lower lobe, 1% in the fissure between the upper and lower lobe and the specific site was unknown in 3% of cases), and 54% were found in the right lung (9% in the right upper lobe, 17% in the right middle lobe, 22% in the right lower lobe, 4% in the fissure between the middle and upper lobe, 1% in the fissure between the middle and lower lobe and the specific site was unknown in 1% of cases) [3] (Figure 4).

Solitary masses or nodules in the lung fields, especially in the right middle lobes, without apparent metastatic lesions, are the most common radiology manifestation [8] (Figure 5).

PSH is easy to be misdiagnosed preoperatively. Cases are seldom diagnosed by performing Computed Tomography (CT)-guided percutaneous lung biopsy because of the insufficient tissue at the puncture site. There are few reports about the PSH developing in the oblique fissure in lungs, with patients bearing such tumors for more than 15 years [4].

**Figure 4:** Multiple pulmonary sclerosing hemangioma in the medial and lateral segments of the middle lobe of the right lung and in the basal segment of the left lower lobe.

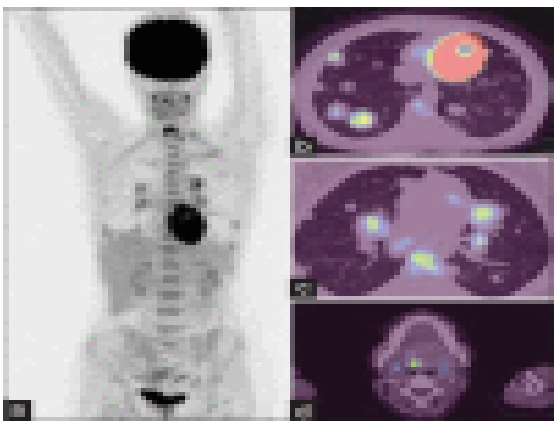

**Figure 5:** Computed tomography scan.

Fluorodeoxyglucose Positron Emission Tomography (FDG PET) has been shown to be more accurate than contrast enhanced CT in differentiating malignant from benign pulmonary nodule. However, false-positive results occur. To the best of our knowledge, only few case reports had reported F18-FDG PET of PSH [9,10]. The co-existence of chronic inflammation along with other common microscopic findings could be a possible factor causing SH to be FDG avid on PET scans [11]. Benign and slow-growing tumors usually showed low glucose metabolism [12]. These larger PSH probably have more active cell proliferation which may lead to higher FDG uptake; or may have more cell components, which are responsible for higher FDG uptake than smaller tumors. Therefore, larger PSH will be misreading as a malignant neoplasm [13]. Previous reports have indicated that, although rapid progression might occur in a solitary PSH, multiple PSHs tend to be slow-growing, another case study described multiple PSHs that were stable over 10 years [14].

PSH can be misdiagnosed as a malignant tumor during intraoperative frozen-section assessment and has a misdiagnosis rate of 25 to 56%, which results in unnecessary extensive surgical procedures and malignant biological behaviors, including local lymph-node metastasis and tumor recurrence [1,15-19].

Definite diagnosis of cases of bilateral multiple PSH is usually possible after postoperative histopathological examination [4]. PSH is mainly composed of two types of cells: cuboidal surface cells that tend to differentiate into type II pneumocytes, and polygonal stromal cells that have considerable multilineage differentiation potential [20]. In the histopathological analysis, both surface-lining cuboidal cells and pale polygonal cells stained positive for thyroid nuclear factor 1, whereas only some of the cells stained positive for vimentin, epithelial membrane antigen, CD68, and cytokeratins [4]. Wang and colleagues suggested that these differences in morphology and phenotype may be attributed to the differences in the status of the cuboidal and polygonal cells [20].

The tumor cells include typical superficial lining cells and underlying round cells, which might origin from the pulmonary epithelial cells, characterized by the positive staining of epithelial markers, such as EMA and TTF-1. But the pathological mechanisms are needed to be further elucidation.

The most meaningful finding of the current immunohistochemical study was the completed positive reaction of EMA and TTF-1 both with the lining cells and round cells. Other epithelial markers, such as CK, CK-7 and PCK, also disclosed full stains with the lining cells [8]. Surgical resection is currently widely used for treating PSH, and no study has reported distant metastasis and the effect of surgical resection on patient survival [4].

Yasushi Adach et al. [21] founded that PSH with lymph node metastasis tended to occur more often in relatively young male patients than PSH without metastasis. The mean size of primary PSHs that had lymph node metastasis was larger than the mean size of non-metastatic primary PSHs [21].

However, PSH predominantly affects middle-aged individuals whose lung function may be bad, and the misdiagnosis rate by intraoperative frozen-section assessment is high, resulting in unnecessary extensive surgical procedures and discomfort for the patients. He et al. suggest that surgical resection should not be considered the preferred strategy for treating single or multiple PSH in the intralobar sites or oblique fissure. If PSH is diagnosed before surgery, and the lesion has no effect on the patient's respiratory function, the patients can survive bearing the tumor [4].

## Conclusion

Most individuals with PSH are asymptomatic or have some non-specific symptoms. Their lesions are usually found accidentally by chest imaging. Although PSH often shows typical imaging characteristics of benign neoplasm of the lung, it is difficult to establish a defined pathological diagnosis preoperatively. The significant error or deferred rate of intraoperative frozen-section evaluation for PSH may result in some unnecessarily extensive surgical procedures. The complete surgical resection is considered the only effective treatment for PSH, and the normal pulmonary tissue should be reserved as possible [2].

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