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Case report

Pulmonary sclerosing pneumocytoma, a rare tumor of the lung

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A B S T R A C T

Pulmonary sclerosing pneumocytoma (PSP) is a rare benign pulmonary tumor. Usually diagnosed incidentally by chest X-ray or chest CT scan. We presented a case of PSP in a 50-year-old woman who was diagnosed with a nodular lesion in the right lung. Thoracotomy was used for the excision of the mass. Pathologic examination revealed no malignant cells. Immunohistochemical studies were performed. TTF-1 was (+), Napsin-A was found to be weakly (+). After surgical resection, the patient was followed up.

1. Introduction

Pulmonary sclerosing pneumocyte (PSP), formerly known as pulmonary sclerosing hemangioma, is a rare pulmonary tumor initially described by Liebow et al., In 1956 as a tumor with marked sclerosis and vascularization [1]. PSP is usually seen in adults over 50 years old, with a female to male ratio of 5: 1 [2,3]. PSP has been shown to be the primitive epithelial origin, most likely from type II alveolar pneumocytes by Immunohistochemical markers [2]. The essential feature of the PSP is the presence of cuboidal surface cells and stromal round cells, both of which are thought to be neoplastic [4]. In the 2015 World Health Organization (WHO) classification, “miscellaneous tumors” have been switched to “adenomas” [5].

We are presenting this PSP case, it is rare, benign disease, might be confused with malignities and there are difficulties in its diagnosis and treatment.

2. Case report

A 50-year-old female patient applied to our clinic in March 2017 after a nodular lesion in her right lung was detected in another hospital. The patient had no complaints, no smoking or tuberculosis history. There was no significant trait in her clinical history. She was not using any medication. Vital findings were followed, as blood pressure arterial 120/70, pulse rate 90/min, respiratory rate 14/min and Oxygen saturation in room air 97%. Her respiratory system examination was normal. WBC 8200, hmg 14.1, Plt 400000, urea 24, creatinine was 0.6. Contrast-enhanced Thorax CT showed a well-defined, hypodense soft-tissue lesion in the size of 28 × 16 mm closely located to the right inferior pulmonary vein (Fig. 1). In PET-CT, a mass of soft tissue with a lobulated contour with no evidence of increased FDG uptake was detected in the superior segment of the right lower lobe of the lungs in the medial paramediastinal area in sizes of 31 × 22 mm. A transthoracic biopsy was planned under the guidance of tomography but the patient refused. Endobronchial pathology was not detected by fiber optic-bronchoscopy. The patient was then referred to thoracic surgery who proceeded to a right lateral thoracotomy. Frozen sample was sent during right lateral thoracotomy. Immunohistochemistry analysis of the resected lesion showed positive (+) with TTF-1, bcl-2 and pansitokeratin, weakly (+) with Napsin-A and CD99 and negative with CD34 (Figs. 2 and 3) consistent with sclerosing pneumocytoma. Sclerosing pneumocytoma was detected after incisional biopsy of the lower lobe of the right lung. Postoperative complications did not develop and the patient was followed-up.

3. Discussion

PSP is a benign tumor with low prevalence. It is often seen in middle-aged Asian women. Patients are usually asymptomatic and it is detected coincidentally. A cough, chest pain and hemoptysis may also occur [6]. Although PSP is often solitary, well-defined, round or oval, homogeneous nodule or mass, there is no definitive diagnostic radiographic finding [7]. However, there are also cases of metastases to the lymph nodes, pleura, and bones [8–10]. Patients may present with a mass lesion of up to 7 cm through 73% of the lesions are below 3 cm [2,4]. Our case had also applied to our clinic with a mass lesion that was detected incidentally. “Air meniscus sign” is a sign for pneumoconiosis [11]. Marginal pseudocapsules (50%), overlying vessels (26.3%), air gap (2.6%) and halo sign (17.1%) are among thoracic CT findings [7]. Sometimes pleural-based, polypoid lesions may mimic a

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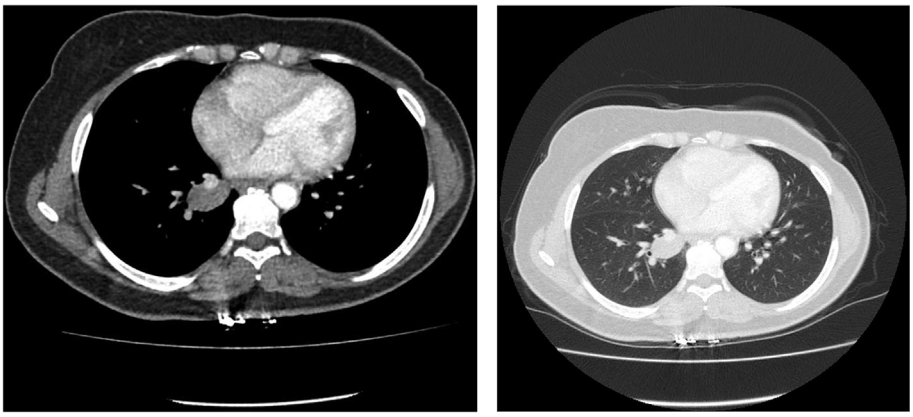

Fig. 1. Contrast-enhanced Thorax CT showed hypodense soft-tissue lesion in the size of 28 × 16 mm closely located to the right inferior pulmonary vein.

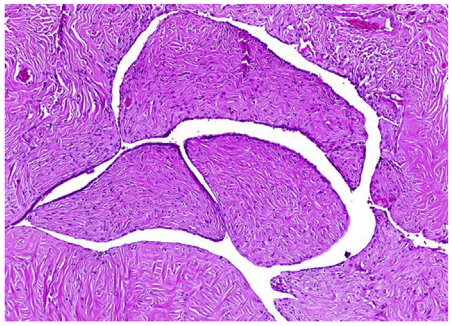

Fig. 2. HE stain shows sclerosing papillary structures are covered by surface cells (50X).

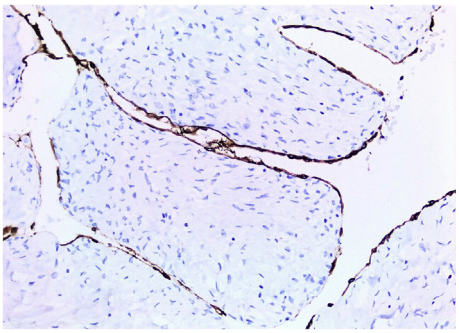

Fig. 3. The surface cells are positive for pancytokeratin and TTF-1 immunohistochemically (100X).

solitary fibrous tumor.

Four typical structural patterns are defined in the PSP; papillary, sclerotic, solid and hemorrhagic. Often the tumor consists of superficial cuboidal and round interstitial cells with a combination of four patterns. If the papillary component of the sclerosing pneumocytoma in the biopsy material is predominant, the diagnosis may be difficult. In addition, both superficial cuboidal cells and round interstitial cells are positively immunoreactive for TTF1 and EMA [2,12]. TTF1 is used in the diagnosis of lung adenocarcinoma and may be misleading for PSP.

Napsin A, a human aspartic proteinase, shows immnohistochemical reactivity in type II pneumocytes with a granular and cytoplasmic staining pattern [13]. It has been widely used in the panel of diagnosis of lung adenocarcinoma along with TTF-1 [14]. Recently it has been demonstrated that Napsin A preferentially stains cuboidal surface cells, not the stromal round cells in sclerosingpneumocytoma [15,16]. The round cells are generally uniformly negative for pan-cytokeratin and positive for cytokeratin-7 and CAM 5.2 in few cases [2]. P53 mutation was exhibited in primary SH. The mutation rate in polygonal cells was higher than that in surface cuboidal cells [17].

The treatment of PSPs is based on surgical resection. Rarely, these tumors can lead to hemoptysis or airway obstruction and respiratory failure [18]. In the study by Park et al. it was determined that wedge resection or enucleation is curative [19]. However, recently, different treatment approaches have been suggested. In a published case report, radiological follow-up decision was taken for a patient diagnosed with PSP by biopsy [20]. It is appropriate that each patient is assessed in a multidisciplinary fashion. Radiotherapy was also suggested as an alternative treatment for inoperable patients [21].

4. Conclusion

PSP is an asymptomatic, rare benign neoplasm. It is mostly incidental. Histologically, it contains two epithelial cell types, surface cells and round cells, which consist of four structural patterns. Preoperative diagnosis is difficult. Surgical excision alone is sufficient for treatment.

Conflicts of interest

The authors have no conflicts of interest relevant to this article to disclose.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.02.002.

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