

PEComa of the Lung: A Rare Entity

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Abstract

Background: Perivascular epithelioid cell tumor (PEComa) is extremely rare neoplasm, especially for the pulmonary localization. Clear-cell tumor of the lung, arising from perivascular epithelioid cells, has mainly asymptomatic and benign course. The diagnosis is confirmed on the immunohistochemical examination. The tumors are usually benign, but there are some sarcoma-like masses. Treatment of choice is a surgical resection. **Case Report:** We report a case of benign PEComa in a 64-year old non-smoking female, accidentally found on a chest X-Ray. VATS anatomic segmentectomy of basal segments was performed. The immunohistochemical examination showed typical features of PEComa. **Conclusion:** PEComa should be considered in the differential diagnosis of pulmonary nodules. Surgical resection is a treatment of choice followed by immunostaining.

Keywords: Fluorescence, Perivascular Epithelioid Cell Neoplasms, Solitary Pulmonary Nodule, Thoracic Surgery.

Introduction

PEComa is a rare tumor of the lung with less than 50 cases reported worldwide. This is a mesenchymal tumor that consists of immunohistochemically distinctive perivascular epithelioid cells [1]. We report a case of asymptomatic PEComa of the right lower lobe and review of the literature.

Case Report

A 64 years old non-smoker female was administered to our hospital due to a right pulmonary nodule, accidentally found on a chest X-ray during a routine examination. Her medical history was unremarkable. Chest CT revealed a 19 mm solitary nodule with well-defined margins in S10 of the right lower lobe [Fig.1a,b]. Abdominal and pelvic CT showed no signs of metastasis. Therefore, video assisted thoracic surgery (VATS) resection of S10 with an express histological examination of the specimen was suggested. A wedge resection

of the lesion was performed. The mass had no visible capsule or signs of pleural invasion; central necrosis and fish-meat like appearance were seen on the cut surface. A frozen section did not exclude malignant lesion. Due to close resection margin and not clear morphologic type of the tumor, the VATS anatomic segmentectomy of basal segments and standard mediastinal lymph node dissection were performed. During operation ICG-fluorescence was used for identification of the inter-segmental plane [Fig.2a,b]. The operation time was 110 min with 15 ml blood loss. Post-operative period was uneventful. The chest drain was removed on a post-operative day two, and patient was discharged without complications.

Histological examination showed structures of the tumor 1.9 cm in diameter, with microcystic transformation, wide myoepithelial septa, clear cell lining, well-defined margins, and perifocal inflammatory component [Fig.3].

Immunohistochemical analysis showed that the tumor had a positive reaction to SMA [Fig.4], CK7, p63 but no reactivity to S100, Melan A, TTF1, RCC, Pax8, Ki67 - 3-5%. Based on these findings, diagnosis of PEComa of the lung was confirmed. There was no evidence of R1 resection or lymph nodes involvement. Adjuvant treatment was not administered, and a regular CT follow-up was recommended [Fig.5a,b].

Discussion

PEComa is a group of rare mesenchymal neoplasms, including angiomyolipoma, lymphangiomyomatosis, clear-cell (sugar) tumor of the lung (CCTL), clear-cell myomelanocytic tumor of falciiform ligament, and clear-cell tumor of other anatomical sites. WHO defines PEComa as a mesenchymal tumor composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [1]. PEComas have unpredictable pathologic behavior, with mostly benign course. These tumors are mostly seen in a gastrointestinal tract and pelvis, with less than 50 pulmonary cases reported in English literature [2]. CCTL was originally described in 1971 by Liebow and Castleman [3]. It is also called “sugar” tumor because of high glycogen concentration in the cytoplasm [4].

CCTL shows no gender predominance and usually occurs in adults in 40-60s [5], however, two reports describe the tumor in a pediatric population [6,7]. But in the paper by J.L. Hornick and C.D. Fletcher PEComas were reported to show a marked female predominance [8]. CCST has been reported in association with LAM, tuberous sclerosis and PEComas of other localization, however, the majority of cases are sporadic [9,10]. Only 7 cases of malignant CCST are reported in the worldwide literature [11]. The factors of malignant potential are large size (>5 cm), infiltrative growth, high nuclear grade and hyper-cellularity, mitoses >1 per 50 hpf, necrosis or vascular invasion [12]. A local recurrence and metastasis to lung, mediastinum,

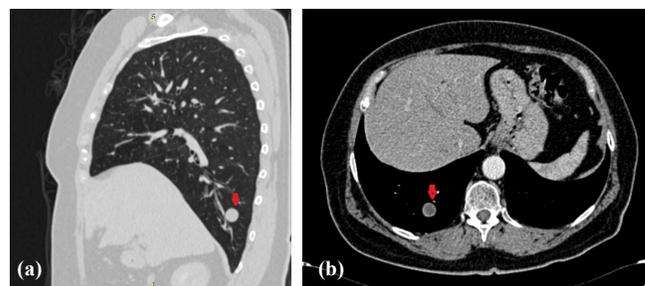


Fig.1(a): A sagittal plane of CT scan in lung window and **(b):** axial plane in mediastinal window showing 19 mm solitary nodule with well-defined margins in S10 of the right lower lobe.

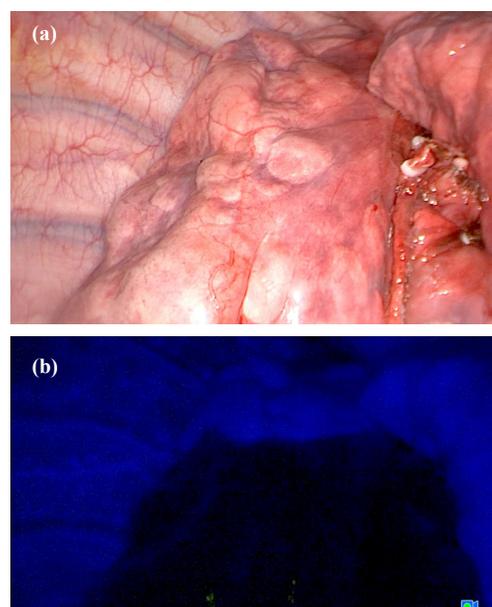


Fig.2(a): An intraoperative image in white light and **(b):** near-infrared mode used to define intersegmental boundaries for right segmentectomy S7-10.

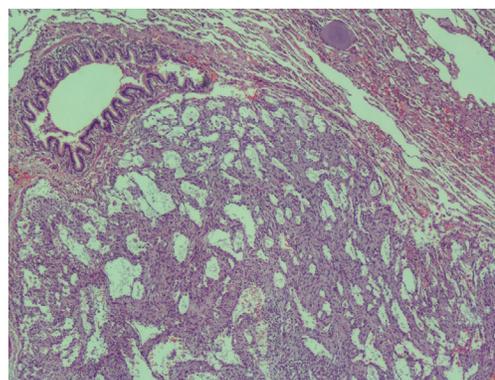


Fig.3: Histological examination of the specimen, showing margins of the tumor.

adrenal gland, temporal lobe and bones are described in case reports [11].

CCTL usually presents as asymptomatic peripheral solitary nodule with no evidence of cavitation or calcification [13]. The typical characteristics of the CCTL on contrast-enhanced CT scans are the intense heterogeneous enhancement in the arterial phase and washout in the delay phase [14]. A single report describes an extensive FDG uptake of the benign tumor [5]. Only two cases presented with hemoptysis [15,16] and one with shortness of breath [17]. Unspecific symptoms are also reported like fever, sputum, fatigue, night sweat, emaciation [4]. S.Sen *et al.* presented a case of PEComa-associated thrombocytosis that decreased after resection of the tumor [2].

Only in one report, a diagnosis was achieved through transbronchial lung biopsy [18]. Transthoracic biopsy under CT navigation is reported to be helpful in differential diagnosis [12, 19], nevertheless, it was not widely advised due to tumor's rich blood flow and features of sinusoidal vessels [2]. An intra-operative fine needle aspiration is a diagnostic option, however, only single reports demonstrate the effectiveness of this method [1]. Thus, the diagnosis is often made only after immunohistochemical analysis of the specimen that shows a positive reaction to HMB-45, SMA and S-100 and no cytokeratin activity [20].

CCTL is often misdiagnosed as metastatic clear cell renal carcinoma, especially during intra-operative cytological examination [4]. A differential diagnosis should be made between primary pulmonary carcinoma, pulmonary metastasis, including metastatic renal cell carcinoma and metastatic melanoma, solitary fibrous tumor, paraganglioma, tuberculoma, granular cell tumors, oncocytoma, acinic cell tumor, sclerosing hemangioma, hemangiopericytoma, and chemodectoma [4,5,21].

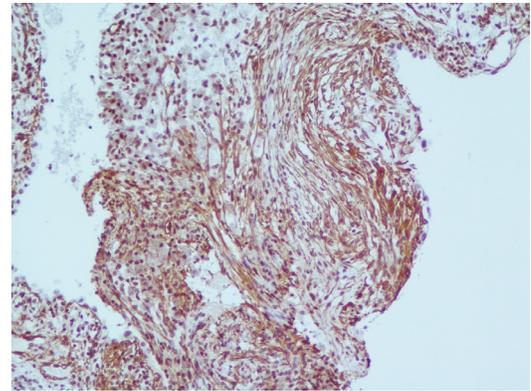


Fig.4: Immunohistochemical analysis of the tumor, showing positive reaction to SMA.

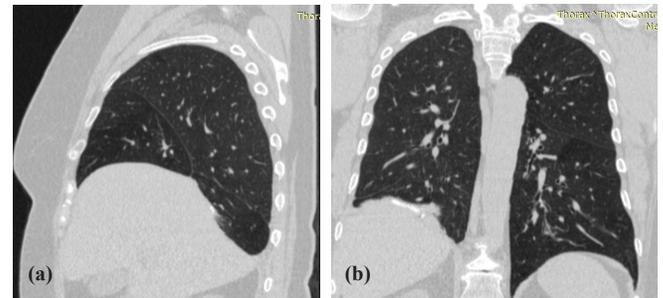


Fig.5: A follow-up chest CT scan in (a): sagittal and (b): coronal planes with no signs of local recurrence.

According to WHO recommendations, lung resection is a treatment of choice and no adjuvant therapy is recommended. A volume of resection is still a matter of debate, including lobectomy, segmentectomy, wedge resection, enucleation, and even pneumonectomy. Recent limited studies reported positive results of targeted therapy administration of mTOR inhibitors in a metastatic PEC tumor of retroperitoneum, kidney, and uterus [11].

Conclusion

This paper highlights the clinico-pathological aspects of pulmonary PEComa with an overview of possible methods of morphologic verification and discussion of lung resection volume. This disease should be considered in a differential diagnosis of solitary pulmonary nodules.

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