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Rare Benign Tumor of the Lung: Perivascular Epithelioid Tumor (Pecoma)

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Case report

History

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ABSTRACT

Aim

Perivascular epithelioid tumors (PEComas) are a group of rare mesenchymal neoplasms, including angiomyolipoma, lymphangioleiomyomatosis, and clear cell tumors of the lung. To date, only around 50 cases have been reported in the literature. Herein, we present a very rare case of pulmonary PEComa to contribute to the existing literature.

Case

A 66-year-old female patient for dyspnea was referred to our clinic. Radiological findings showed a nodular mass lesion in the lung. Laboratory findings were within normal limits. In the thorax computed tomography, a 34*33 mm mass lesion was resected. Histopathological examination reported it as 'clear cell tumor-PEComa.

Conclusion

Given the unpredictable behavior of these tumors, we recommend anatomical resection in cases where the intraoperative frozen section does not clearly distinguish between benign and malignant pathology.

***Keywords*:** *Perivascular epithelioid cell neoplasms, PEComatous tumors, Lung, Lung neoplasms*

Akciğerin Nadir Benign Tümörü: Perivasküler Epiteloid Tümör (Pecoma)

Olgu sunumu

Süreç

Geliş: 05/10/2022
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ÖZ

Amaç

Perivasküler epiteloid tümörler (PEComa), anjiyomiyolipom, lenfanjiyomiyomatozis ve akciğerin berrak hücreli tümörlerini içeren bir grup mezenkimal tümördür. Litaratürde şu ana kadar sadece 50 vaka bildirilmiştir. Literatüre katkıda bulunmak amacıyla akciğerin çok nadir görülen perivasküler epiteloid tümörünü sunmayı amaçladık.

Vaka

66 yaşında kadın hasta dispne nedeniyle kliniğimize sevk edildi. Radyolojik incelemede sağ akciğerde nodüler kitle saptandı. Laboratuvar bulguları normal sınırlardaydı. Toraks bilgisayarlı tomografisinde, 34*33 mm'lik bir kitle lezyonu çıkarıldı. Histopatolojik inceleme, 'berrak hücreli tümör-PEComa' olarak bildirildi.

Sonuç

Bu tümörlerin gelecekteki davranışı net olarak bilinmediğinden, 'dondurulmuş kesit'te malign/benign ayrımı yapılamayan vakalarda bu hastalar için anatomik rezeksiyon öneriyoruz.

Anahtar kelimeler: Perivasküler epiteloid hücreli neoplazileri, PEComatöz tümorler, Akciğer, Akciğer neoplazmları

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Introduction

Benign lung tumors account for less than 1% of all pulmonary neoplasms. Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms characterized by unique histological and immunohistochemical features. These tumors typically express both melanocytic and smooth muscle markers and have been documented in various anatomical locations including the lung, kidney, and soft tissues. To date, approximately 50 pulmonary PEComa cases have been reported in the literature.[1]

Diagnosing PEComa can be challenging due to its histological resemblance to other neoplasms. Accurate diagnosis often requires comprehensive histological, immunohistochemical, and sometimes molecular analysis. In this case, we report a rare pulmonary PEComa detected in a 66-year-old female patient who underwent a middle lobectomy.

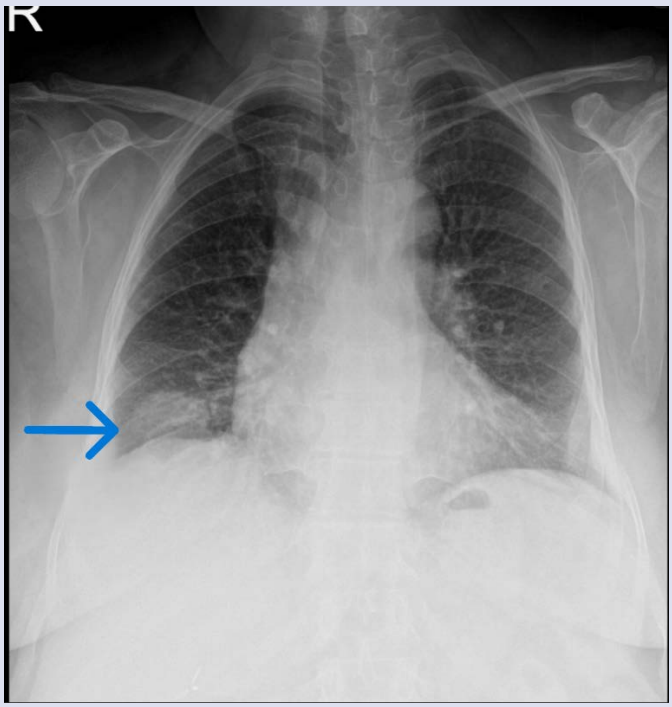


Figure 1: Preoperative posterior anterior chest radiograph of the patient

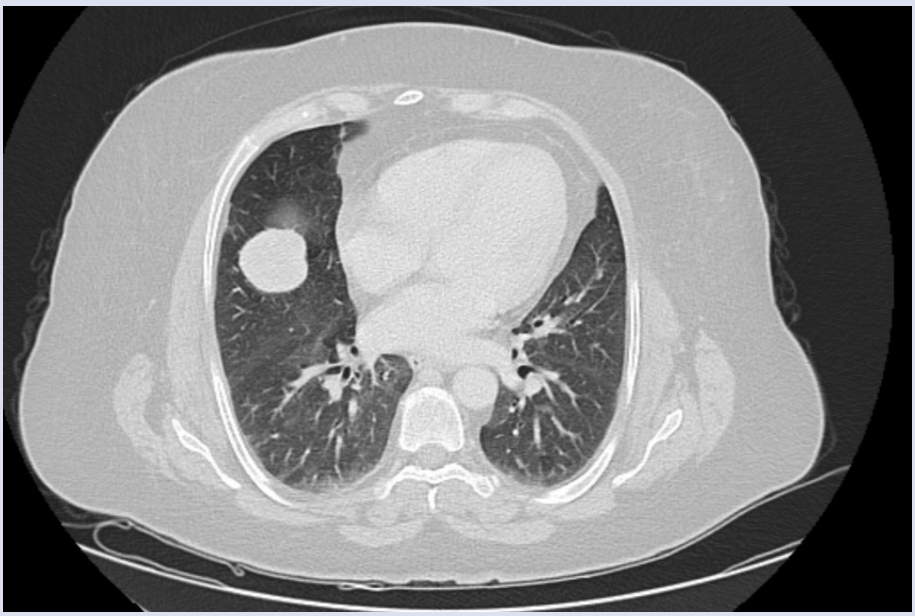


Figure 2: Thoracic CT image of the patient

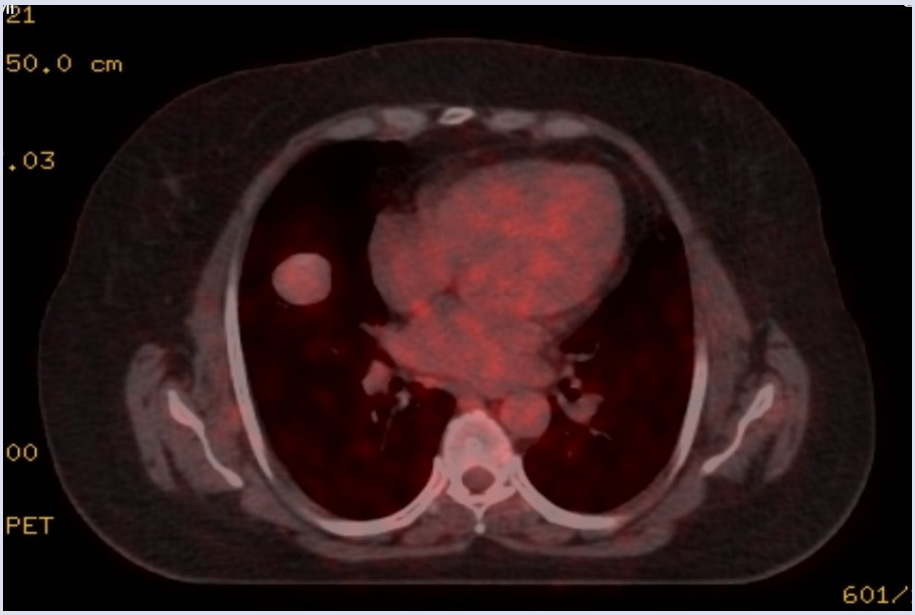


Figure 3: Pet-CT image of the patient

Case

A 66-year-old woman was referred to our clinic for evaluation of dyspnea. Imaging revealed a nodular mass in the lung. Laboratory findings were within normal ranges. She had no significant comorbidities. Pulmonary function testing revealed an FEV1 of 1.83 liters.

A posterior-anterior chest radiograph demonstrated a radio-opacity in the lower zone of the right lung (Figure 1). Thoracic CT showed a 34 × 33 mm mass located in the lateral segment of the middle lobe of the right lung, adjacent to the diaphragmatic pleura (Figure 2). Comparison with a CT scan performed one year earlier revealed no change in the size of the lesion. 18F-fluorodeoxyglucose (FDG) PET-CT demonstrated increased uptake in the same region (SUVmax: 2.9) (Figure 3).

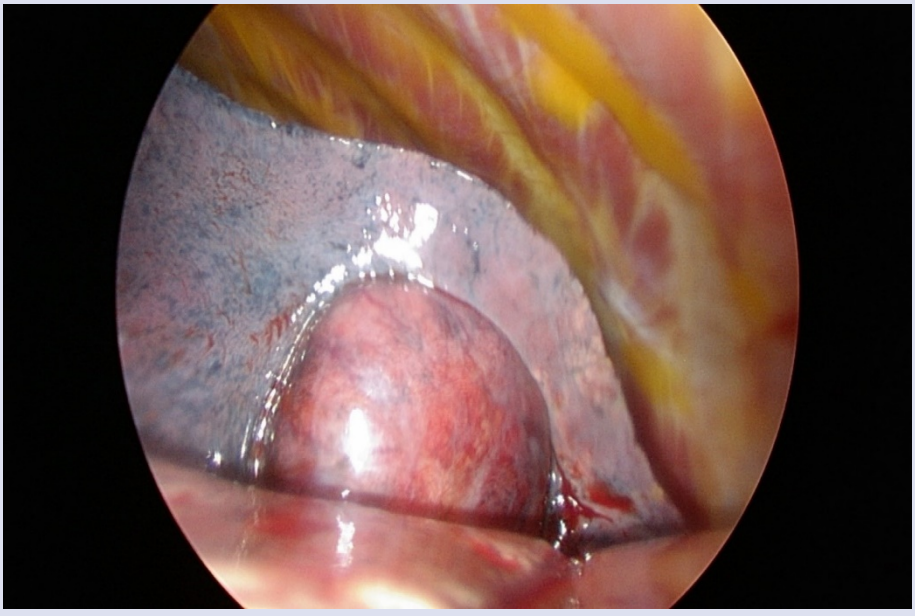


Figure 4: Intraoperative view of PEComa

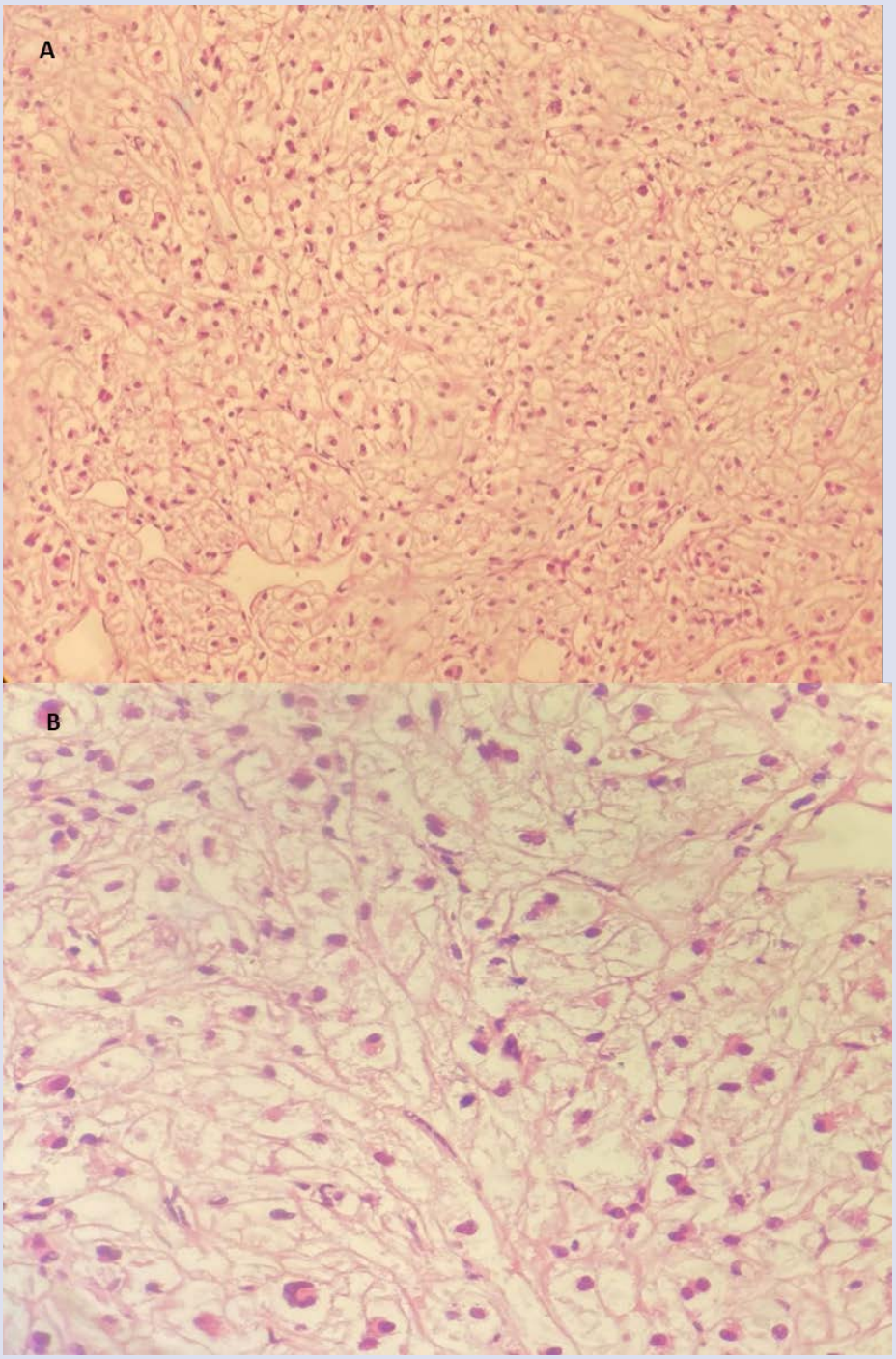


Figure 5: A . A diffusely developed tumor composed of epithelial cells without any pattern is observed (HEx200).
Figure 5: B. It is noteworthy that the tumor cells are large, slightly pleomorphic, with hyperchromatic nuclei and clear cytoplasm (HEx400).

A frozen section analysis was performed using samples obtained via video-assisted thoracoscopic surgery (VATS) (Figure 4). However, due to the inability to definitively classify the lesion as benign or malignant, a middle lobectomy with mediastinal lymph node dissection was conducted. Histopathological examination revealed a clear cell tumor consistent with PEComa (Figure 5).

Microscopically, the tumor exhibited a nested and sheet-like architecture, composed of uniform round to ovoid cells with abundant clear cytoplasm and well-defined borders. Tumor cells were associated with blood vessel walls and were arranged radially around thin-walled vessels, often showing subendothelial growth. Nuclear atypia was mild, with absent mitotic figures. Immunohistochemical markers for melanocytic and smooth muscle differentiation were negative.

The patient was discharged on the eighth postoperative day without complications. She has been followed for 45 months with no evidence of recurrence or metastasis.

Results and Discussion

Although non-small cell lung carcinoma is the most prevalent type of lung cancer, PEComas represent an extremely rare subset of pulmonary tumors.[2] When located in the lung, they are referred to as clear cell tumors or "sugar" tumors. The first such case was described by Liebow and Castleman in 1963. These tumors predominantly affect females and can occur at any age between 8 and 73 years.[3] Only over 50 cases have been reported in the literature.[1]

PEComas are often incidentally detected as solitary pulmonary nodules, generally measuring less than 3 cm in diameter. However, it was seen as a mass lesion in our case.[4] Tumor size greater than 2.2 cm is considered a potential risk factor for malignancy. In this case, the tumor measured 3.4 cm.[5]

Frozen section analysis was performed intraoperatively; however, the nature of the tumor could not be definitively determined. Consequently, middle lobectomy and mediastinal lymph node dissection were carried out. Histologically, PEComas consist of round, oval, or polygonal cells with distinct borders and abundant clear or eosinophilic cytoplasm rich in glycogen.[6]

Pulmonary PEComas may be misdiagnosed as metastatic clear cell renal carcinoma, especially in frozen sections. Definitive diagnosis often relies on a thorough histological and immunohistochemical assessment.[7] To date, eight cases of malignant pulmonary PEComas have been reported. Due to their rarity, no consensus exists regarding their optimal management.[3]

Conclusion

In conclusion, due to the uncertain long-term behavior of pulmonary PEComas, we recommend anatomical resection in cases where frozen section analysis is inconclusive. This approach allows for both definitive diagnosis and potential therapeutic benefit.[8]

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