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*CLINICAL CASE*

# Atypical presentation of an unusual lung tumor

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## Summary

*We describe a case of a rare bronchial epithelial-myoepithelial carcinoma of the lung in a 36-year-old man. Tumor enucleation was carried out and the patient was put on long-term follow up. Tumor categorisation was difficult. The patient is disease-free on the 30th postoperative month. The clinicopathological uniqueness of this neoplasm is reviewed.*

**Key words:** epithelial, immunohistochemical, lung, myoepithelial, salivary gland, tumor

## Introduction

Epithelial-myoepithelial tumors are most frequently encountered in the salivary glands, and in a relatively low incidence in the sweat glands or in the breast [1]. Salivary gland-type tumors of the lung are rare, despite the widespread distribution of mucosal-submucosal glands in the tracheobronchial tree [2].

The biological behavior, clinical outcome, prevalence, the spectrum and the malignant potential of these tumors are not clearly understood [3]. Generally, they behave as low-grade malignant neoplasms [2,3]. Commonly, lobectomy, bronchial sleeve resection, bilobectomy or even pneumonectomy are the treatments of choice [1,3-6].

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## Case presentation

A 36-year-old man was admitted with sweating, weakness, and exertional dyspnea of one month duration. Both chest radiograph and computed tomography scan showed a solid right lower lobe mass (Figure 1). An endobronchial growth pattern was seen at bronchoscopy. A hydatid cyst was diagnosed clinically and further endoscopic biopsy was avoided. An exploratory thoracotomy was performed to provide definite diagnosis. Frozen section was considered

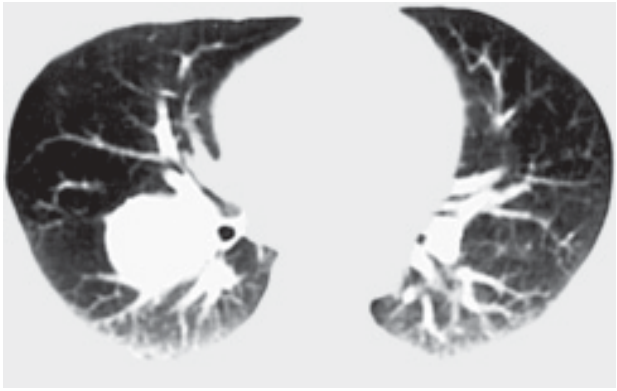

**Figure 1.** Computed tomography revealing a 4X5X5 cm round mass at the right lower lobe in close contact with the lower lobe bronchus.

benign, so enucleation of the mass was carried out. The tumor was dissected from the surrounding parenchyma but the bronchial communication was rigid, suggesting bronchogenic origin of the tumor. The bronchus was perforated and repaired. No further intra- nor postoperative complications were detected and the patient was discharged on the 8th postoperative day.

Diagnostic conflicts appeared during pathologic examination of the tumor. First, it was assumed to be a metastasis from testicular cancer, then it was considered as a large cell carcinoma with rhabdoid foci (Figure 2), and finally it was accepted as an epithelial-myoepithelial carcinoma. The final diagnosis took more than one year. During this period, although the tumor was characterized malignant, we decided to put the patient on follow up instead of performing a completion lobectomy. The patient remains with no evidence of disease recurrence on the 30th postoperative month (Figure 3).

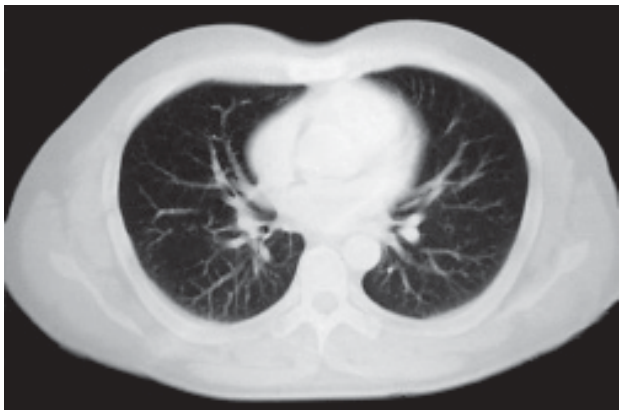

**Figure 3.** Computed tomography of the same level on the 30th postoperative month shows local fibrotic changes at the operation site. Neither local recurrence nor metastasis was discovered.

## Discussion

Myoepithelial neoplasms mainly occur in the salivary glands and breast, being extremely rare in the lung. The most common tumors of the tracheobronchial glands include adenoid cystic carcinoma, mucoepidermoid carcinoma, pleomorphic adenoma, and other rare tumors including oncocytoma and acinic cell carcinoma [1,7]. Pulmonary neoplasms composed mainly or exclusively of epithelial and myoepithelial cells have been designated in the past under a variety of descriptive terms, including adenomyoepithelioma, epithelial-myoepithelial tumor, and epithelial-myoepithelial carcinoma [1]. Recently, it has been proposed to name these tumors as pulmonary epithelial-myoepithelial tumors of unproven malignant potential (PEMTUMP) [2].

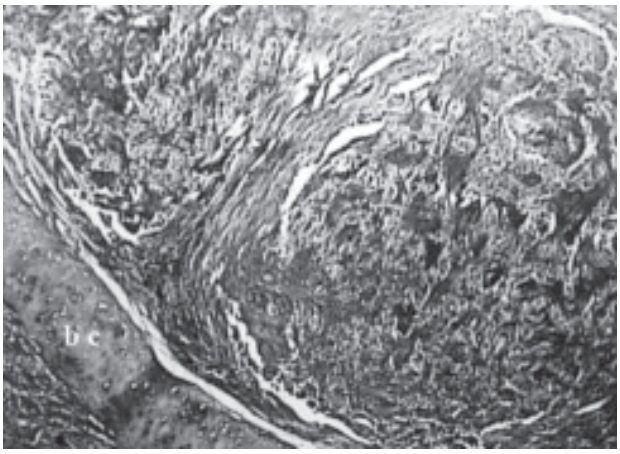


**Figure 2.** Photomicrograph of the specimen shows ill-defined lobules of tumor cells in a fibrous stroma. Bronchial cartilage also seen at the left lower corner possibly implies that the tumor originates from the lung (H&E X25).

These tumors are usually polypoid in shape and commonly located in the upper airways [2,7]. Eight cases with a lower respiratory tract tumor have been reported [2, 6], and there is only one case in the literature without any connection with the bronchial tree [8]. Tumors in the tracheobronchial tree give symptoms relatively early, usually when they reach 2 cm in diameter. In contrast, neoplasms that extend into the lung parenchyma do not give symptoms in some cases before they reach up to even 16 cm in diameter [7]. In our patient, the tumor was 5 cm in diameter, originated from the lower respiratory tree and grew into the parenchyma.

Epithelial-myoepithelial tumors of the salivary glands behave as low-grade malignant neoplasms [1,2,7,9]. The interval between the initial diagnosis and local recurrence is approximately 5 years. Distant metastases may occur even 15 years later [2]. The follow up period of the reported cases range from 6 to 36 months [6]. Our follow up period was 30 months and we have not yet detected local recurrence or distant metastases (Figure 3).

The diagnosis of myoepithelial tumors of the lung, especially the determination of subgroups, is difficult [10]. At the beginning, this case was thought to be a testicular metastasis, because the patient was male. Some areas consisted of groups of uniform

tissue separated by fibrous septa. Following immunohistochemistry, it was suggested to be large cell carcinoma with rhabdoid foci (Figure 2). Further immunohistochemical evaluation showed a tumor consisting of confluent clusters of cells in areas divided by fibrous septa. The cellular preservation was suboptimal but they appeared to have a clear cytoplasm. No mucin or mucous glands were identified. The tumor cells were cytokeratin (AE1/3) positive, but they were negative for CK7, TTF-1 and SMA (smooth muscle actin). Classification of this tumor was difficult, but it was thought to represent a primary pulmonary salivary gland neoplasm. The patient's psychologic status was worsened by the delay of diagnosis. We suggested the patient that he should be on close follow up in order not to miss the correct time for a complementary operation in case this would be necessary.

Parenchyma-saving resection is proposed for these type of tumors because of their low-grade malignant behavior. However, it is difficult to decide on the appropriate surgical therapy perioperatively without a definite frozen section diagnosis. Such tumors are difficult to diagnose on paraffin sections and it would be wrong to give a definite diagnosis on frozen section. The reported cases provide an accumulation of clinical, pathological, and particularly prognostic information about this rare entity.

## References


1. Higashiyama M, Kodama K, Yokouchi H et al. Myoepithelioma of the lung: report of two cases and review of the literature. Lung Cancer 1998; 20: 47-56.
2. Pelosi G, Fragetta F, Maffini F, Solli F, Viale G. Pulmonary epithelial-myoepithelial tumor of unproven malignant potential: report of a case and review of the literature. Mod Pathol 2001; 14: 521-526.
3. Wilson RW, Moran CA. Epithelial-myoepithelial carcinoma of the lung: immunohistochemical and ultrastructural observations and review of the literature. Hum Pathol 1997; 8: 631-635.
4. Miura K, Harada H, Aiba S, Tsutui Y. Myoepithelial carcinoma of the lung arising from bronchial submucosa. Am J Surg Pathol 2000; 24: 1300-1304.
5. Mori M, Furuya K, Kimura T et al. Mixed tumor of salivary gland type arising in the bronchus. Ann Thorac Surg 1991; 52: 1322-1324.
6. Doganay L, Bilgi S, Ozdil A, Yoruk Y, Altaner S, Kutlu K. Epithelial-myoepithelial carcinoma of the lung. Arch Pathol Lab Med 2003; 127: 177-180.
7. Moran CA, Suster S, Askin FB, Koss MN. Benign and malignant salivary gland-type mixed tumors of the lung. Clinicopathologic and immunohistochemical study of eight cases. Cancer 1994; 73: 2481-2490.
8. Strickler JG, Hegstrom J, Thomas MJ. Myoepithelioma of the lung. Pathol Lab Med 1987; 111: 1082-1085.
9. Ryska A, Kerekes Z, Hozorkova E, Borton P. Epithelial-myoepithelial carcinoma of the bronchus. Pathol Res Pract 1998; 194: 431-435.
10. Morans CA. Primary salivary gland-type tumors of the lung. Semin Diagn Pathol 1995; 12: 106-122.