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## Pilomatrix Carcinoma with Subsequent Pulmonary Metastases: a Case Report

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### ABSTRACT

*Pilomatrix carcinoma is the malignant counterpart of the pilomatrixoma which is a hamartoma of the hair matrix. It is a rare locally aggressive skin tumor with a great tendency for recurrence but its metastatic potential is limited.*

*We report a pilomatrix carcinoma with proven metastases to the lung in a 51-year old man who was admitted to the hospital for respiratory symptoms such as productive cough with green colored sputum and halitosis since one year ago. (Tanaffos 2006; 5(3): 57-60)*

**Keywords:** Pilomatrix carcinoma, Metastasis, Lung, Skin

### INTRODUCTION

Pilomatrix carcinoma is the malignant counterpart of the pilomatrixoma which is a hamartoma of the hair matrix (1). It is a rare locally aggressive skin tumor with a great tendency for recurrence but its metastatic potential is limited.

The tumor can metastasize to distant organs such as the lungs, bone and lymph nodes. At least four cases with visceral metastases occurring several years after the primary diagnosis have been reported.

Here, we report a pilomatrix carcinoma with proven metastases to the lung.

### CASE SUMMARIES

A 51-year-old man was admitted to Ghaem Hospital on August 2005 for respiratory symptoms such as productive cough with green-colored sputum and halitosis since one year ago. He also reported a mild vague pain in his left hemithorax and nonspecific complaints including weight loss, anorexia and intermittent fever.

The review of his past medical history revealed that the patient had a subcutaneous mass in his back since childhood. This nodule was constant in size for many years but started a sudden accelerated growth three years ago. Thus, the patient sought for treatment and the tumor was excised by a general surgeon. Six months later a tiny mass reappeared at the previous site and reached its primary size in two

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months. A secondary excision was performed but it recurred again. This time the lesion showed a more aggressive behavior with invasion to skeletal muscles; in this session the lesion was completely removed with an adequate safe margin and from that time, the skin tumor did not recur.

The patient did not have anything remarkable in his medical history.

A chest x-ray showed a mass in the left lung (2) (Figure 1).

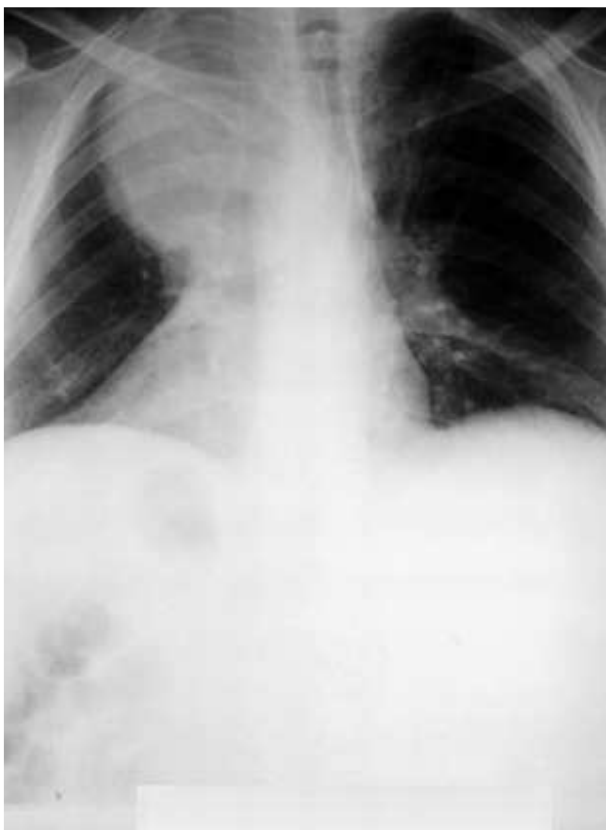


Figure 1. Chest x-ray of patient.

CT-scan demonstrated a large mass in upper regions of left hemithorax with coarse calcification foci that caused the left upper lobe to collapse (3) (Figure 2).

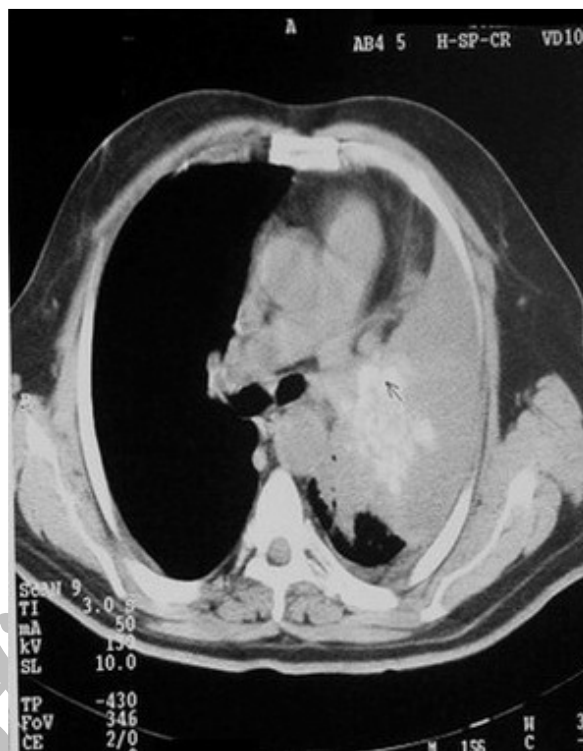


Figure 2. Thoracic CT-scan

The spirometry pattern was obstructive. Blood chemistry was normal and complete blood count testing showed the following findings:

Hb=11.3 g/dl, WBC count= 15.9, RBC count=4.1  
Hct=35 %, Lymph= 9 % , PMN %=84%

Bronchoscopy demonstrated a vegetative lesion with necrotic surface at the entry of left upper bronchus which caused an obstruction there. Hemorrhage occurred during lavage.

The pathologic examination of the endobronchial biopsy and histologic evaluation of the cutaneous lesion previously excised, revealed a dense infiltrate of basaloid cells, an abrupt transition to shadow cells, and central necrosis. "Malignant Pilomatrixoma" was reported (Figure 3, 4)

Then both left pneumonectomy and pericardiectomy were performed. (Figure 5).

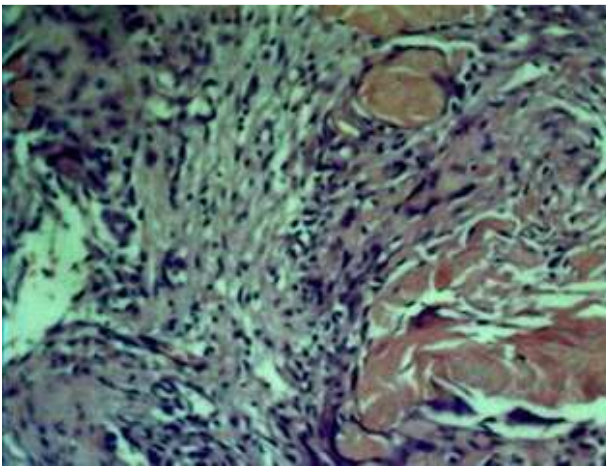


Figure 3. Pulmonary lesion (H&E X400)

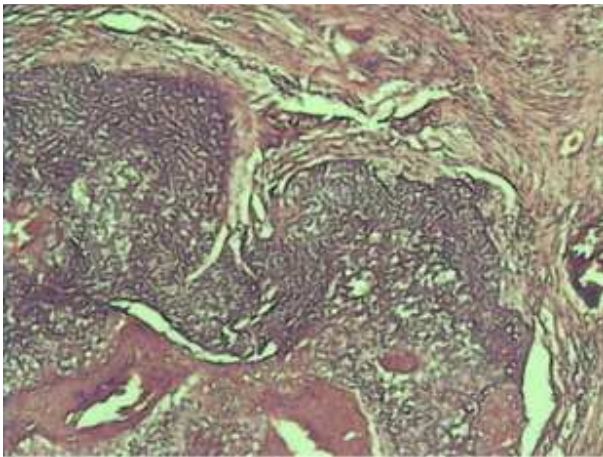


Figure 4. Cutaneous lesion (H&E X200)



Figure 5. Left pneumonectomy

## DISCUSSION

Pilomatrix carcinoma is a very rare skin tumor (4). Around 70 cases are reported in the literature. The lesion has a male predominance and occurs in older patients in contrast to pilomatrixoma which occurs mostly in females under the age of 20. Some pilomatrixomas show apparent transformation into carcinomas. Other cases are malignant from the onset. The tumor presents as a rapidly expanding firm nodule and can cause metastases (5, 6, 7, 8).

From the histopathologic point of view, pilomatrix carcinomas are asymmetrical, cellular, infiltrative neoplasms. Many areas especially the periphery of the tumor show proliferations of large, anaplastic, hyperchromatic basophilic cells with numerous mitoses; however, this can be seen in pilomatrixomas on occasion (9). Toward the center of the tumor there may be transformation of basophilic cells into eosinophilic shadow cells, seen in benign pilomatrixomas, or there may be large cystic centers containing necrotic debris. Features which are helpful in making the diagnosis include asymmetry and poor circumscription, presence of several markedly sized and variably shaped basaloid aggregations of tumor cells, continuity of basaloid cells with the epidermis, extensive areas of necrosis, infiltrative growth pattern, and presence of ulceration.

Both the primary tumor and its metastasis reveal extremely high proliferation and apoptotic rates (10, 11). High constant expressions of CD44v6 and P-cadherin are also observed. The best approach for assessment of metastatic potential of pilomatrix carcinoma seems to be the complex evaluation of routine histological criteria like vessel invasion, mitotic index, apoptotic count, and new molecular markers of cell death and adhesion (12). The differentiation of these tumors from benign pilomatrixomas depends on a constellation of microscopic features, some of which may be equivocal or absent in individual biopsy specimens.

On 2002, De Galvez et al. reported a pilomatrix carcinoma on the right knee. The tumor developed at

the site of a previous lesion that had been present for 28 years. Histologic study showed the presence of basaloid cells with numerous atypical mitoses, shadow cells and calcification. After several surgical excisions, the tumor mass infiltrated the subcutaneous tissue, muscle and bone resulting in inguinal lymph node and pulmonary metastases.(13)

Bremnes and his colleagues in 1999 presented a 74-year-old male with a pilomatrix carcinoma from the left temporal region developed in July 1996 which was excised. One month after diagnosis, metastases to both lungs and to a regional lymph node were found and histologically verified. The patient also developed metastases in the abdomen, back and thoracic spine. The latter resulted in spinal cord compression and paraplegia.(14)

In 1996, another case of pilomatrix carcinoma with multiple visceral metastases was reported.(15)

Gould et al. reported the first metastasizing pilomatrix carcinoma in 1984. Their patient was a 67-year-old male with locally recurrent pilomatrix carcinoma of the back that caused bilateral pulmonary metastases within a 4-year period (5).

One can observe that almost all of the reported cases of viscerally metastatic tumors involved the lungs mostly bilaterally (16, 17).

In general, because of the possibility of the metastatic spread of the pilomatrix carcinoma, correct diagnosis and wide local excision of the tumor are required and follow up is recommended to identify possible metastases (18).

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