

## Pilomatrix Carcinoma with Lung and Lymph Node Metastases

Pilomatrix carcinoma is a rare locally aggressive hair-follicle tumor. We report a 54-year-old man who presented with a tumor in the left flank that was found by skin biopsy to be pilomatrix carcinoma. A contrast-enhanced computed tomographic scan of the chest, abdomen, and pelvis showed multiple small nodules in both lungs and lymphadenopathy in the abdomen. Video-assisted thoracoscopic biopsy of the lung lesions was consistent with metastatic pilomatrix carcinoma. After intravenous cisplatin and 5-fluorouracil, the skin, lung, and lymph node lesions shrank. (*J Lung Cancer* 2008;7(2):90–92)

**Key Words:** Pilomatrix carcinoma, Multiple metastases, Chemotherapy

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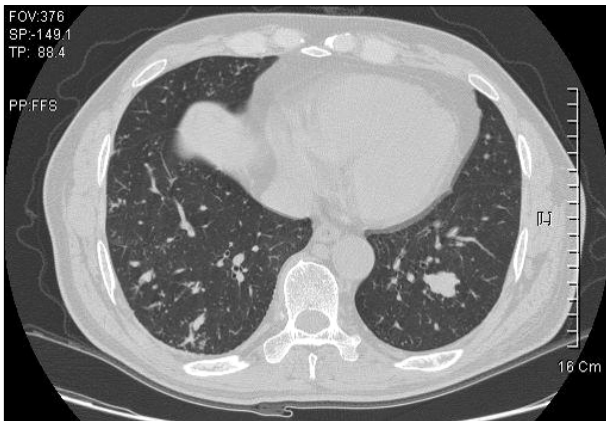
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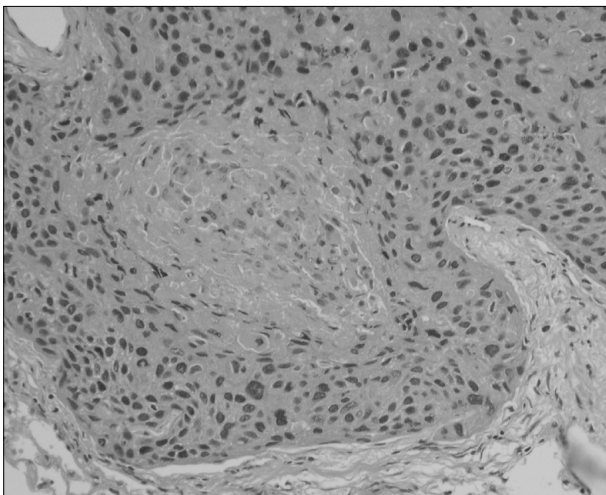
Although Malherbe and Chenantais first described pilomatrixoma in 1880 as a “calcifying epithelioma,” the disorder was not recognized for several decades as a neoplasm in which malignant transformation could occur. Gromiko (1) reported a case of an aggressive and recurrent pilomatrixoma in 1927, and the use of the designation “pilomatrix carcinoma” or “calcifying epitheliocarcinoma of Malherbe” was proposed by Lopansri and Mihm (2) in 1980, when the investigators presented a case of aggressive pilomatrixoma with a review of five similar cases from the literature. However, despite the local aggressiveness of the tumor and a tendency to recur, the presence of a distant metastasis is rare. We present a case of a pilomatrix carcinoma in a 54-year-old man with lung and lymph node metastases that were responsive to systemic chemotherapy.

### CASE REPORT

A 54-year-old man presented with a tumor in the left flank, which had been first noticed five years prior. The tumor had been growing rapidly for a year, and the patient was referred to the Department of Dermatology of our hospital. The patient had smoked a pack of cigarettes per day for the previous 30 years and had a history of occasional alcohol abuse. The patient had sustained a left humeral fracture 20 years prior. His mother had hypertension. On a physical examination, the patient appeared well. The temperature was 36.2°C, the pulse rate was 74 beats/min, the respiratory rate was 20/min, and blood pressure was 130/90 mmHg. On the left flank, a 13×15 cm skin mass elevated 1 cm above the skin surface was found. A chest radiograph revealed reticulonodular opacities on both lower lung zones. A contrast-enhanced computed tomographic (CT)



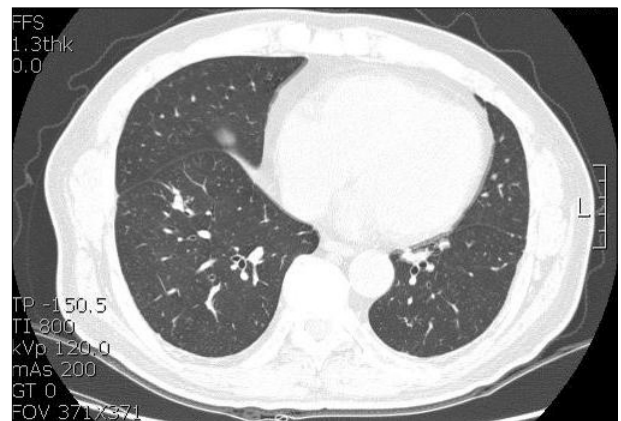
**Fig. 1.** Initial chest CT scan showing branching opacities with tree-in-bud appearance consistent with tortuous pulmonary arteriolar dilatation by tumor embolism in both lungs.



**Fig. 2.** Lung biopsy showing a nest of basaloid tumor cells with hyperchromatic, pleomorphic nuclei and small distinct nucleoli and necrotic or shadow cells in the center (Hematoxylin-eosin,  $\times 100$ ).

scan of the chest showed multiple lesions suggestive of lung metastases and enlargement of axillary lymph nodes (Fig. 1). A CT scan of the abdominal and pelvis showed multiple areas of lymphadenopathy that were likewise suggestive of metastases.

A biopsy of the tumor in the left flank and a video-assisted thoracoscopic biopsy of one of the lung lesions were performed. A histological evaluation demonstrated the presence of nests of basaloid cells with hyperchromatic and pleomorphic nuclei and prominent nucleoli in the dermis and epidermis. In the center of the tumor, transitional zones with retained nuclei



**Fig. 3.** Follow-up chest CT scan after four courses of cisplatin and 5-fluorouracil demonstrating improvement of lung lesions.

and shadow cells were seen. The lung specimen was consistent with a metastasis from a pilomatrix carcinoma (Fig. 2). Immunohistochemical staining for high-molecular weight cytokeratin (34bE12) and epithelial membrane antigen were positive. Staining for carcinoembryonic antigen was focally positive and staining for S-100 was weakly positive. Staining for low-molecular weight cytokeratin 7 was negative. Six courses of chemotherapy with intravenous cisplatin (100 mg/m<sup>2</sup> on day 1) and 5-fluorouracil (1,000 mg/m<sup>2</sup>/24h on days 1~5) were given. Follow-up CT scans of the chest and abdomen after the second and fourth cycles showed shrinkage of the lung and lymph node metastases (Fig. 3). The patient has continued to visit for follow-up two years after the initial diagnosis.

## DISCUSSION

Approximately 80 cases of pilomatrix carcinoma have been reported, most of which have shown locally aggressive behavior with a tendency for recurrence. However, the metastatic potential is thought to be limited, and only a few cases with metastases to visceral organs, bone, and lymph node have been described to date(3-8). A pilomatrix carcinoma has a predilection for the head and neck region, especially for the posterior neck and upper back, as in the case of a pilomatrixoma. Unlike the benign form, a pilomatrix carcinoma occurs predominantly in male and older patients(7). The major clinical concern may be in the differentiation of this rare malignant neoplasm from the more frequent benign form, which histologically resemble each other. An examination of pilomatrixomas shows bands of

basaloid cells with small uniform nuclei and scanty pale cytoplasm, usually at the periphery, and organized areas of keratinization with a shadow or ghost cells in the center(9). Both immunohistochemical and flow cytometric analyses have been performed to distinguish a pilomatrix carcinoma from its benign counterpart; however, neither of these methods has been successful(10). Therefore, the histological features still provide the most important clues for diagnosis. The main indicators of malignancy are nuclear pleomorphism, frequent and atypical mitoses, central necrosis, infiltration of the skin and soft tissue, vascular invasion and ulceration(11). Differentiation from other tumors such as a basal cell carcinoma with matrical differentiation, trichoepithelioma, lymphoepithelioma-like carcinoma of the skin, squamous-cell carcinoma, and mixed tumors of the skin is necessary(3).

Once a pilomatrix carcinoma is diagnosed histologically, further evaluation including liver function tests, a serum calcium assay and chest radiography should be performed. A CT scan or MRI examination needs to be performed when aggressive local invasion is suspected(12). The local aggressiveness of the tumor and the likelihood of recurrence make wide excision with histologically confirmed negative margins advisable. Wide local excision decreases the local recurrence rate, which can be more than 50% after simple excision(7). Adjuvant radiation therapy has also been used(2). In the case of metastatic disease, chemotherapy and radiation have been given. To the best of our knowledge, the use of systemic chemotherapy has been attempted in three cases of metastatic pilomatrix carcinoma, and none of the cases showed a response (4-6). In the present case, a follow-up CT scan of the chest and abdomen after four courses of cisplatin and 5-fluorouracil documented the shrinkage of the skin lesion and partial

response of the metastatic lung and lymph node lesions.

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