

## Metastatic Angiosarcoma of the Lung : HRCT Findings<sup>1</sup>

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We describe a case of cavitory metastasis to the lungs from a small angiosarcoma of the scalp, in which the metastatic lesions were complicated by pneumothorax and pulmonary hemorrhage. On high-resolution CT, the lesions simulated the findings of Langerhans cell histiocytosis. Thin-walled cavitory metastatic lesions were similar to those of thin walled air cysts in Langerhans cell histiocytosis. Ground-glass opacity simulated the findings of smoker's respiratory bronchiolitis in Langerhans cell histiocytosis but histologically represented hemorrhage during metastasis of the angiosarcoma.

**Index words :** Neoplasms, metastases

Sarcoma

Pneumothorax

Lung, CT

Computed tomography (CT), high-resolution

Angiosarcoma is a rare malignant tumor of the skin and soft tissue, and commonly metastasizes to the lungs, showing multiple nodules. The metastatic lung lesions rarely cavitate(1-5), though a angiosarcoma with cavitory metastasis can be complicated by hemopneumothorax(1-5). Metastatic lung lesions showing both cavities and pulmonary hemorrhage on high resolution CT have not, however, been reported. We describe a case of angiosarcoma of the scalp which rapidly developed cavitory lung metastasis, bilateral pneumothoraces, and diffuse pulmonary hemorrhage, and led to the patient's death.

### Case Report

An 85-year-old non-smoking man was admitted with dyspnea; its onset had been sudden, it persisted for a week and was accompanied by a cough, fresh hemoptysis and chest pain. Fifteen days before admission he had undergone excision biopsy of a soft tissue mass in the parietal area of the scalp. The histologic findings of the lesion were consistent with angiosarcoma involving the dermis and subcutaneous fat.

The results of a pulmonary function test, performed on admission were normal. Cytologic examination of the sputum revealed no malignant cells, and sputum culture showed no bacterial growth.

Initial chest radiograph showed increased interstitial markings and numerous thin-walled cavitory lesions in both lungs, small amount of pneumothorax was seen in the left pleural space. The chest radiograph obtained six days later showed an area of ground-glass opacity in the

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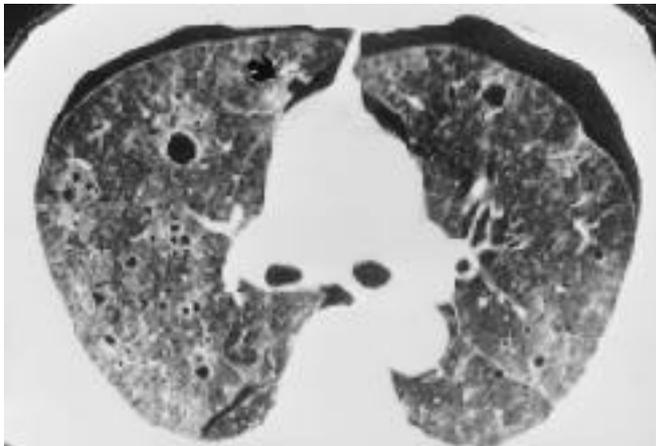
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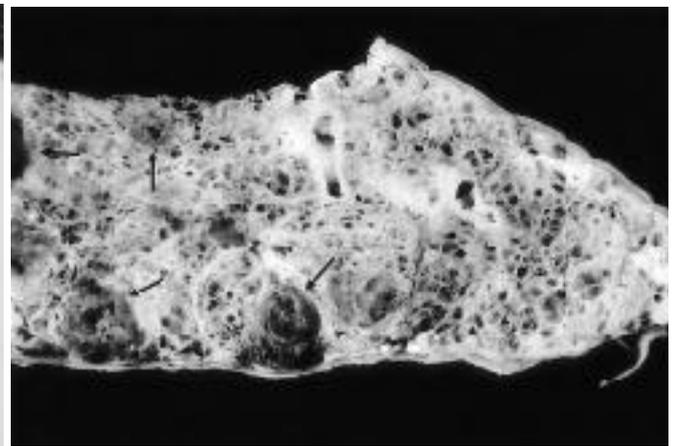
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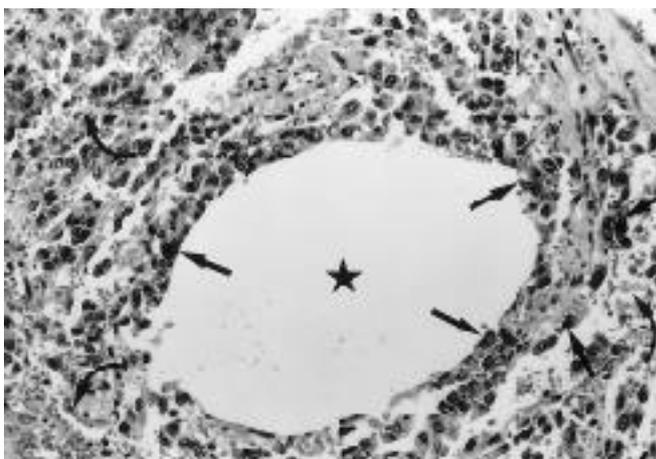
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Fig. 1. 85-year-old-man with pulmonary metastasis from an-  
giosarcoma of the scalp presenting with abrupt onset of dyspnea  
and hemoptysis for 1-week.

A. Initial chest radiograph shows left pneumothorax and multi-  
ple cavities predominantly in the both upper lungs (arrows).

B. Follow-up chest radiograph taken on 2- weeks after (A) shows  
bilateral pneumothoraces even with chest-tube drainage and in-  
creased number of cavities and extent of ground-glass opacity.  
Note also extensive bilateral subcutaneous emphysema.

C. High-resolution CT at that time of A shows innumerable cavi-  
ties and bilateral pneumothoraces. Although many cavities ap-  
pear round, they also had bizarre shapes, about 3-13mm in size,  
with distinct wall, mimicking cysts of Langerhans cell histiocy-  
tosis. A pleural-based cavity is partly opened into pleural space  
(arrow), representing a rupture site. There are multifocal areas  
of ground-glass opacity in the both lungs.

D. Cut surface of the resected lung shows hemorrhagic nodules (arrows) with central cavitory change.

E. Microscopically, one hemorrhagic nodule shows central cavitory change (star) and tumor shows large, plump cells (arrows)  
forming wall of cavity and RBC filled vascular spaces (curved arrows) (H-E stain,  $\times 200$ ).

right upper lobe and findings of new pneumothorax in the right pleural space. On high-resolution CT(1 mm collimation, 140kVp, 170mA, 512x512 matrix, a bone algorithm, lung window width of 1500H and window level of 700), cavitory lesions of 3-13mm in diameter were distributed evenly throughout both lungs. The cavities were mostly round but the shape of some was bizarre. In the walls of cavities there was no discernible nodule at the component, but diffuse ground-glass opacities were present in both lungs. After admission, cavities, pulmonary hemorrhage, subcutaneous emphysema and pneumothoraces with air leakage became much more extensive, and the patient's general condition progressively deteriorated until he died of pulmonary hemorrhage and respiratory failure on the ten days after admission. Necropsy was performed and lung tissue was obtained from the right lower lobe.

Grossly, resected lung tissue was diffusely hemorrhagic and contained many cysts measuring 3 - 13 mm in diameter. The histologic features of lung lesions were similar to those of the scalp lesion, indicating that the former were metastatic angiosarcoma. They were however, more anaplastic and hypervascular than the scalp lesion. The cavities were mostly empty, through some contained blood clots and exudates. Many areas showed perivascular invasion, and in some cysts, all lining cells were neoplastic. No vessels contained tumor emboli.

### Discussion

The term Angiosarcomas encompasses formerly called hemangiosarcomas and lymphangiosarcomas. An angiosarcoma originate from the endothelial cells and arises most commonly from the dermis of the scalp and upper half of the facial area(6), and is frequently accompanied by pulmonary metastasis in which solid nodules or masses are produced. Thin-walled cavernous pulmonary metastases are rarely reported(1). Generally, neoplastic cavities occur in approximately nine percent of primary and four percent of metastatic pulmonary tumors(2).

In the case we describe, the pulmonary metastatic lesions were unusual in three ways : in almost all metastatic nodules there was thin-walled cavitation, diffuse ground-glass opacity on high-resolution CT, and bilateral pneumothoraces, presumably caused by the rupture of subpleural cavities, and which led to the patient's death. The radiologic findings of diffuse bizarre shaped cystic lesions, predominantly in the upper lobes, and as-

sociated with bilateral pneumothoraces, mimicked the radiologic findings in cases of Langerhans cell histiocytosis. Because the patient's clinical setting precluded infection, rapidly evolving ground-glass opacities were initially regarded as re-expansion edema caused by decompression of pneumothorax or atelectasis caused by compression. The cause of ground-glass opacity was pulmonary hemorrhage(Fig. 1. C). Histopathologic examinations of both resected lung(Fig. 1 D, E) and scalp mass showed diffuse hemorrhage and hemosiderin-laden macrophages, with frequent perivascular invasion of tumor cells.

The pathogenesis of cavitory pulmonary metastasis is controversial. It may be caused by tumor necrosis secondary to inadequate blood supply due to tumor emboli and subsequent evacuation of necrotic tissue into the airway, a mechanism which can explain the pathogenesis of rapidly growing large lesions. The cavitory lesions in the present case, however, were small and on histological examination showed no tumor emboli. As an alternative, the tumor, by extrinsic compression, can cause bronchial obstruction and thus a resulting cystic lesion, along which tumor cells can extend to form tumor cavities. Nomura et al.(4) suggested that in angiosarcomas, the lining of the vessels tends to be incomplete and that these imperfect vessels can break down and coalesce to form thin-walled cavities.

In the present case, cavitory metastasis was complicated by bilateral pneumothorax and subsequently by soft tissue emphysema resulting from continuous air leak. A few subpleural cavities were deformed and their walls were disrupted, they were presumed to be the sites of origin of associated pneumothorax. Dines et al.(8) suggested that bronchopleural fistula is a pathogenic mechanism of pneumothorax in metastatic sarcomas, the fistula being produced by rupture of the peripheral necrotic lesions into both the bronchial tree and pleural space. They also suggested that the lung parenchymal infarction distal to tumor emboli might be responsible for air-leak into the pleural space.

The case described in this paper illustrates a rare form of cavitory metastasis to the lungs from a small angiosarcoma of the scalp that was complicated by pneumothorax and pulmonary hemorrhage. The high-resolution CT findings simulates the findings of Langerhans cell histiocytosis.

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