Angiosarcoma with Pulmonary Metastasis Presenting with Spontaneous Bilateral Pneumothorax in an Elderly Man

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Cutaneous angiosarcoma is a rare and invasive endothelial-derived sarcoma that occurs most frequently in the scalp and facial skin of elderly men. It is frequently accompanied by thin-walled cavitary pulmonary metastasis, and is often obscure on chest radiograph. We report a case of angiosarcoma of the scalp with cystic metastasis to the lung in a 63-year-old man, presenting as recurrent bilateral spontaneous pneumothorax. Lung metastasis was missed at the first episode of pneumothorax because the lung-expanded chest radiograph showed no significant abnormality. Two months later, bilateral pneumothorax recurred, and high-resolution computed tomography revealed multiple cystic, cavitary and nodular lesions. Pulmonary metastasis was confirmed by video-assisted thoracoscopic surgery, and pleurodesis was performed. After the operation, the patient received chemotherapy; no recurrence of pneumothorax was found during 6 months of follow-up. Pneumothorax in the elderly should be differentiated from malignant metastatic lung tumors. [J Formos Med Assoc 2006;105(3):238–241]

Key Words: angiosarcoma, cystic lung metastasis, pneumothorax

Angiosarcoma is a rare malignant tumor that originates from endothelial cells, and has an extremely poor prognosis. Angiosarcoma is frequently accompanied by pulmonary metastasis with presentation of thin-walled cavitary metastasis. We report a patient with angiosarcoma of the scalp who suffered recurrent bilateral spontaneous pneumothorax, which was missed at the first episode because the lung-expanded chest radiograph showed no significant abnormality.

Case Report

A 63-year-old man was treated at the emergency department after sudden onset of shortness of breath for 2 hours. He had been admitted to another tertiary care hospital 4 months prior to this admission because of an erythematous nodule on his forehead. Total excision of the skin tumor had been performed, followed by skin grafting and adjuvant radiotherapy after the pathologic diagnosis of angiosarcoma. Two months prior to this admission, he had been re-admitted to the same hospital due to sudden onset of dyspnea. Bilateral pneumothorax was found, and closed thoracostomy was immediately performed for the treatment of pneumothorax. He was discharged after a chest radiograph showed full expansion of both lungs and no specific abnormal findings.

On this admission, he presented to our emergency department with severe dyspnea for 2 hours.
Physical examination revealed a well-nourished man in acute respiratory distress with accessory muscle use. Chest auscultation revealed bilaterally diminished breath sounds, but no heart murmurs were heard. He was afebrile, with a blood pressure of 106/65 mmHg, a pulse of 110 bpm, and a respiratory rate of 29 breaths/min. According to the previous medical history and physical findings, bilateral pneumothorax was tentatively diagnosed (Figure 1) and emergency thoracostomy was immediately performed on the right hemithorax. Laboratory data were significant for a white blood cell count of 11,600/μL, hemoglobin of 19.6 g/dL, and platelet count of 252,000/μL. Other laboratory data included the following: alanine aminotransferase, 52 IU/L; blood urea nitrogen, 19 mg/dL; creatinine, 1.0 mg/dL; glucose, 125 mg/dL. He was subsequently admitted to our hospital for further management.

Although both lungs fully expanded with continuous chest tube drainage, air leakage persisted on the 4th hospital day. A high-resolution computed tomography (CT) scan of the chest (Figure 2) revealed multiple cystic, cavitary and nodular lesions in both lungs, subcutaneous emphysema, and persistent pneumothorax despite chest tube drainage. Video-assisted thoracoscopic surgery revealed disseminated small pinkish nodular indurations over both parietal and visceral pleura with profound vascularization and anthracosis. All the visible nodules seemed fragile and showed easy-contact bleeding during forceps clamping. A 45-mm endoGIA stapling device (US Surgical Corp, Norwalk, CT, USA) was used to remove the pleural nodules, which were immediately diagnosed pathologically by frozen section as metastatic angiosarcoma. Air leak leading to pneumothorax was also checked by artificial ventilation of the lung, after immersing the lung in warm saline solution, which showed bubbles produced from pleural tear of the metastatic nodules. Multiple wedge resections were performed to prevent further recurrence of pneumothorax.

Microscopically, a highly vascular tumor characterized by irregularly infiltrative, anastomosing vascular channels lined by pleomorphic cells with tufts was seen (Figure 3A). Immunohistochemical stain showed positivity for CD34 focally (Figure 3B). Histologic findings were compatible with metastatic angiosarcoma.
After the operation, the patient received chemotherapy at another hospital, and had no recurrence of pneumothorax during 6 months of follow-up.

Discussion

Cutaneous angiosarcoma is a rare and invasive endothelial-derived sarcoma that occurs most frequently in the scalp and face skin of elderly men.\textsuperscript{1,2} It tends to recur locally and to metastasize despite aggressive therapy. The lung is the most common site of metastatic involvement, followed by the liver, cervical lymph nodes, spleen, and, rarely, the heart and brain. The mean survival time after metastases was 4 months.\textsuperscript{3} Morgan et al reported that tumor diameter, depth of invasion, positive margins, metastases, and tumor recurrence were the most robust predictors of outcome.\textsuperscript{3}

Pulmonary metastasis of angiosarcoma commonly appeared as extensive solid nodules, but cystic or cavitary pulmonary lesions have also been reported.\textsuperscript{4} The most common presenting symptom of pulmonary metastasis in angiosarcoma was hemoptysis,\textsuperscript{5} while relatively few cases presented with spontaneous pneumothorax, pneumomediastinum or pulmonary hemorrhage.\textsuperscript{6-8} Our patient’s initial presentation was bilateral spontaneous pneumothorax, but chest radiograph with the expanded lung showed no significant abnormality. After the recurrence of pneumothorax, chest CT was performed and detected multiple thin-walled cystic lesions and nodules. Various mechanisms have been proposed for the pathogenesis of thin-walled cavity formation, but no definite conclusions have been reached.\textsuperscript{8} These include excavation of a nodule tumor through discharge of the necrotic material inside and infiltration of malignant cells into the walls of a pre-existing pulmonary bulla. CT scan of the chest in this case demonstrated nodular, cavitary and cystic lesions, which could represent the three stages of tumor progression.

Rare pulmonary diseases could contribute to simultaneous bilateral pneumothorax, such as sarcoidosis, histiocytosis X, lymphangioleiomyomatosis and bullous lung disease.\textsuperscript{9} Lung cancer and other malignancies with pulmonary metastasis usually cause unilateral spontaneous pneumothorax. The mechanism of pneumothorax development in pulmonary metastasis of angiosarcoma could be the result of rupture of peripheral malignant cysts into the pleural space.\textsuperscript{10} In our patient, both CT scan and operative findings demonstrated nodular, cavitary and cystic lesions, which resulted in pneumothorax.

Treatments for cutaneous angiosarcoma are individualized on the basis of extent of disease, anatomic location, and patient consent. Management includes surgical excision alone, adjuvant radiotherapy and chemotherapy. Even with aggressive treatment, the prognosis of cutaneous angiosarcoma is still poor, with a 5-year survival of 10–35%.

Cutaneous angiosarcoma with cystic pulmonary metastasis is rare; moreover, simultaneous bilateral pneumothorax is an extremely rare clinical
Metastatic angiosarcoma of the lung presenting with pneumothorax

presentation. High-resolution CT scan should be helpful in the survey of pulmonary lesions, especially in elderly patients who have underlying lung disease with uncommon presentations.

References