Pneumothorax due to Hemangiopericytoma Metastasis from the Thigh

A 25-year-old female had malignant hemangiopericytoma of the right thigh resected. Three years later, right pneumothorax developed, necessitating a thoracotomy. A 3-cm bulla was discovered on the lower right pulmonary lobe and was subsequently resected. Microscopy showed tumor cells which were positive for actin, but negative for epithelial membrane antigen, vimentin, and factor VIII-related antigen—features compatible with those of the thigh. The patient has been free of the disease for five years. Pertinent literature reveals similar cases with hemangioendothelioma but offers few reports on pneumothorax caused by metastatic hemangiopericytoma. (JJTCVS 1998; 46: 523–525)

Index words: Pneumothorax, hemangiopericytoma, metastasis

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To the best of our knowledge, metastatic hemangiopericytoma of the lungs manifested by pneumothorax has only been reported in French literature.1 Malignant hemangiopericytoma in general carries a poor prognosis in the presence of a pulmonary metastasis.2–4 We have, however, treated a young woman who had undergone resection of hemangiopericytoma in the thigh three years prior to the onset of pneumothorax due to its metastasis. She has been free of the disease since the operation on the pulmonary lesion four years ago, which is herein reported.

Case

A 25-year-old non-smoking female diagnosed with a 7.5 × 7.0-cm tumor in the right anterior thigh, which was resected on September 10, 1990. Microscopy revealed the tumor to be malignant hemangiopericytoma. The patient then received a wide resection of the rectus femoris on September 25, 1990. Histopathology performed on the specimen showed no residual tumor.

In May 1993, right pneumothorax was demonstrated, resulting in sudden pain developing in the ipsilateral chest. She had a chest tube inserted and the pneumothorax subsided. In August 1993, a similar episode arose when a chest X-ray showed right pneumothorax in the absence of emphysema (Fig. 1). A lateral thoracotomy was performed on September 1, 1993, revealing serosanguineous effusion which was cytologically negative. A 3-cm bulla was found on the lower pulmonary lobe facing the interlobar fissure. A pin-size hole on the lesion was found causative of air leakage. The lesion was resected en bloc with part of the lower lobe of the lung. For five years thereafter, the patient was continuously found to be free of the disease.

Photomicroscopy performed on the bullous lesion of the right lung showed tumor cells proliferating in the subpleural space. Hematoxylin-eosin and sil-
Fig. 1. Chest roentgenogram at admission showing right pneumothorax accompanied by some ipsilateral pleural effusion.

Fig. 2. Low-power photomicroscopy of the perforated bullous lesion, hematoxylin-eosin (x20). Up: toward the visceral pleura; down: toward the pulmonary parenchyma; arrow: the area with tumor cells. The tumor proliferates in the subpleural layer and contains an area of hemorrhage.

Fig. 3. High-power photomicroscopy of the tumor, hematoxylin-eosin (x100). Spindle-shaped tumor cells surround blood vessels with irregular luminal patterns.

Discussion

To the best of our knowledge, one case has been reported elsewhere in literature pertaining to pneumothorax caused by malignant hemangiopericytoma metastasis from an extrathoracic organ. A similar entity, malignant hemangioendothelioma, occasionally metastasizes to the lungs. One piece of Japanese literature notes that 39 (48%) of 82 cases of skin-originated hemangioendothelioma showed metastasis to the lungs; 24 (92%) of 26 fully-documented cases had pneumo- and/or hemothorax develop. Such instances of pneumothorax has been attributed to (1) a tumor in the pleura or the bronchiole developing necrosis leading into bronchopleural fistula; and/or (2) the check valve mechanisms facilitating expansion of part of the lung peripheral to the metastasis. In contrast to these mechanisms of pneumothorax in metastatic sarcoma, bronchogenic carcinoma in smokers with bullous emphysema can create pneumothorax by the obstruction and eventual rupture of the pre-existing bulla. Histopathology in our non-smoking patient demonstrated a rupture of the intact pleura and no tumor necrosis, thus indicating that the ball valve mechanism had been operative in the absence of emphysema.

As regards the prognosis of hemangiopericytoma, 16 (15%) of 71 patients showed metastasis to a distant organ within an interval one year to 14 years; two thirds of them were preceded by a local recurrence of the tumor. The 10-year actuarial sur-