Pulmonary artery hypertension of acute onset with an essentially normal chest roentgenogram in a dyspneic patient suggests pulmonary thromboembolism, but can be caused by carcinomatous lymphangitis, which is more difficult to diagnose.

Case Report

A 41-year-old woman was admitted to hospital with severe dyspnea that had begun as exertional dyspnea and palpitations 1 week before admission, and she had had a persistent cough for 2 months. On examination the patient was dyspneic and diaphoretic. Her weight, which was 77 kg, had decreased from 84 kg 1 month previously in association with anorexia. She was 155 cm in height. The pulse rate was 100 beats/min and the blood pressure was 145/90 mmHg. There were no rales over the lung field. Pitting edema of the legs was not present. An electrocardiogram showed sinus tachycardia (101 beats/min), Q waves in lead III, and inverted T waves in leads V1–4 and III, and aVF. Analysis of arterial blood gases sampled while breathing room air indicated a PaO2 of 54.2 mmHg, PaCO2 of 31.3 mmHg, and a pH of 7.46. Hemoglobin was 14.5 g/dl; hematocrit, 45.6%; erythrocyte count, 5,540,000/mm3; and leukocyte count, 7,800/mm3. Lactate dehydrogenase activity was 600 IU/L; fibrinogen, 351 mg/dl; and fibrin and fibrinogen degradation products, 54.4 μg/ml. Prothrombin time was 12.0 s with a control of 16.4 s. A chest roentgenogram was unremarkable except for mild cardiomegaly with a slightly enlarged right pulmonary artery. Echocardiography showed a reduced left ventricle and interventricular septal flattening.

Pulmonary thromboembolism was suspected and the patient was treated with oxygen, urokinase and heparin, which resulted in a slight improvement in her condition. Computed tomography performed the next day revealed dilated left and right main pulmonary arteries, and also enlarged pretracheal lymph nodes. Diffusely distributed fine reticulonodular changes were present in the lung, and a left breast mass was suggested. Perfusion lung scanning showed equivocal defects in the both lungs. Pulmonary arteriography was performed on the third hospital day; the pulmonary arteries appeared normal, although pulmonary arterial pressure was 81/40 mmHg. The dyspnea progressed, and on hospital day 4 the patient was intubated and transbronchial lung biopsy was performed, from which histology diagnosed carcinomatous lymphangitis: small foci of adenocarcinoma were present within lymphatic channels (Fig 1). The cancer cells formed microtubules and had large hyperchromatic nuclei, and rich foamy or eosinophilic cytoplasm. Organized thrombi were observed in small vessels. The patient died of respiratory failure 7 days after hospital admission. Autopsy was not performed.

Discussion

Computed tomography performed the next day revealed dilated left and right main pulmonary arteries, and also enlarged pretracheal lymph nodes. Diffusely distributed fine reticulonodular changes were present in the lung, and a left breast mass was suggested. Perfusion lung scanning showed equivocal defects in the both lungs. Pulmonary arteriography was performed on the third hospital day; the pulmonary arteries appeared normal, although pulmonary arterial pressure was 81/40 mmHg. The dyspnea progressed, and on hospital day 4 the patient was intubated and transbronchial lung biopsy was performed, from which histology diagnosed carcinomatous lymphangitis: small foci of adenocarcinoma were present within lymphatic channels (Fig 1). The cancer cells formed microtubules and had large hyperchromatic nuclei, and rich foamy or eosinophilic cytoplasm. Organized thrombi were observed in small vessels. The patient died of respiratory failure 7 days after hospital admission. Autopsy was not performed.
roentgenogram in the present patient and 2-dimensional echocardiography suggested acute pulmonary artery hypertension. The patient’s condition worsened rapidly, but the etiology was difficult to determine. In a previously reported case examined at autopsy, pulmonary hypertension of acute onset was found to have resulted from carcinomatous lymphangitis representing dissemination of an unsuspected gastric cancer. In the present case, the transbronchial lung biopsy specimen revealed that the pulmonary hypertension was caused by carcinomatous lymphangitis with additional organized thrombi in the small vessels. Although we could not clarify why pulmonary microvascular thrombosis was associated with carcinomatous lymphangitis, obstruction of small vessels by thrombi can be a consequence of pulmonary hypertension. Although Soares et al reported that arrest of arterial tumor emboli could play an important role in pulmonary hypertension, no arterial tumor embolism was observed in our small histological specimen. Organized thrombi are considered to develop from pulmonary endarteritis caused by cancer cells directly infiltrating the arterial walls from the adventitial layer, or by hematogenous spread. The present adenocarcinoma had a nonspecific histologic appearance that was not useful for identifying the primary site of the cancer, although we suspect it arose in the breast, based on the computed tomography findings. Harold found the stomach, breast, lung and prostate to be the most frequent primary sites in an analysis of 178 cases of carcinomatous lymphangitis.

All etiologies should be considered when evaluating a patient with pulmonary hypertension. In the present case, the clinical presentation of carcinomatous lymphangitis was very similar to that of acute pulmonary thromboembolism and so occult carcinoma should be included in the differential diagnosis. We found transbronchial lung biopsy to be very useful for etiologic diagnosis of pulmonary hypertension.

References