A 43-year-old woman experienced increasing dyspnea over a period of 1 year. She was examined first at an outside hospital. Diagnostic workup revealed an elevated precapillary mean pulmonary arterial pressure of >44 mm Hg. Left ventricular function was nearly normal (cardiac index, 2.1 L/min·m²; pulmonary wedge pressure, 15 mm Hg). Computed tomography of the lung demonstrated regional ground glass opacities and pleural effusions but no signs of pulmonary embolism. Perfusion and ventilation scintigraphy showed perfusion defects typical for peripheral lung embolism (Figure 1). The diagnosis of chronic thromboembolic pulmonary hypertension was made. There was no correlate in pulmonary angiography for surgical intervention. Therapy with riociguat was initiated but had to be stopped because of systemic hypotension. Repeat drainage of the pleural effusion resulted in recurrence. Permanent drainage was instituted.

The patient was subsequently referred to our hospital for further evaluation. A new computed tomography of the lung demonstrated fields of ground glass opacities, pronounced mediastinal lymph nodes, interlobular lines, and areas of pulmonary consolidation (Figures 2 and 3). Bronchoalveolar lavage contained siderophages. Cryptogenic organizing pneumonia was considered because of peripheral pulmonary consolidations and no signs of an infection. Therapy with systemic steroids was started. Treatment showed no effect on the disease. Other causes for pulmonary hypertension were ruled out.

The combination of high pulmonary arterial pressure, enlarged mediastinal lymph nodes, and morphological changes of the lung structure with ground glass opacities and interlobular lines was suggestive of pulmonary veno-occlusive disease. As an unusual finding, computed tomography also showed lung consolidation. No medical therapeutic option was available, so the patient was planned for lung transplantation. Unfortunately, the patient died of right-sided heart failure before an organ was allocated. Autopsy confirmed the diagnosis of pulmonary veno-occlusive disease (Figure 4).

Pulmonary veno-occlusive disease is a rare disease with the frequency of 0.1 to 0.2 per 1,000,000.1 Perfusion defects are described in 7% of the patients,2 and only a few cases were reported in detail.3 This case points out that abnormal lung scintigraphy may suggest a false diagnosis of chronic thromboembolic pulmonary hypertension while missing the correct diagnosis of pulmonary veno-occlusive disease. The distinction is important because of completely different treatments and the relative contraindication for pulmonary vasodilators in pulmonary veno-occlusive disease.

Disclosures

None.

References

Figure 3. Computed tomography also demonstrated pleural effusion (asterisk), ground glass opacities (small black arrows), and interlobular thickening (arrowheads). These hallmarks are often detected in patients with pulmonary veno-occlusive disease. However, lung consolidations (black arrows) are an unusual finding.

Figure 4. Pulmonary venule with subtotal luminal occlusion and marked intimal thickening resulting from loose fibrous remodeling. The adjacent lung tissue shows an increase in hemosiderin-laden macrophages (elastica van Gieson, ×400).
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