Case Report

Lung Involvement in Angiotropic Lymphoma: CT Findings

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Angiotropic lymphoma, a rare malignant neoplasm characterized by intravascular proliferation of lymphoma cells, usually affects the skin, the brain, and sometimes the lungs. Lung involvement can be associated with severe dyspnea resulting in death. However, radiologic manifestations of lung involvement have not been well described. To our knowledge, this report is the first describing the CT manifestations of lung involvement in angiotropic lymphoma.

Case Report

A 59-year-old woman presented with a persistent dry cough and low-grade fever. Chest radiography showed diffuse reticulonodular opacities (Fig. 1A). High-resolution CT revealed panlobular ground-glass opacities with weblike intralobular interstitial thickening and possible narrowing of the pulmonary veins (Fig. 1B). No hilar or mediastinal lymphadenopathy was present. A transbronchial lung biopsy specimen was nondiagnostic. The patient’s condition improved under corticosteroid therapy (prednisolone, 40 mg/day), and she was discharged. Her clinical symptoms, however, gradually worsened. She was readmitted to our hospital 2 months later. CT of the chest revealed widespread ground-glass opacities and segmental consolidation with air bronchograms (Fig. 1C). The patient underwent corticosteroid pulse therapy combined with cyclophosphamide administration. However, she died of respiratory failure without improvement of clinical symptoms. Autopsy specimens showed intravascular proliferation of lymphoma cells with minimal extravascular infiltration or moderate fibrotic change (Fig. 1D) and a focal area of dense fibrosis. Evidence of infarction suggested pulmonary emboli. Alveolar hemorrhage was minimal. Immunohistochemical stains proved the tumor to be a B-cell lymphoma. Tumor was seen not only in the lungs but also in the skin, liver, spleen, and kidneys. The adrenals were not enlarged. However, a few lymphoma cells were present in the adrenal capillaries.

Discussion

Angiotropic lymphoma is a rare malignancy characterized by intravascular proliferation of mononuclear cells within relatively small vessels [1]. The condition was first described in 1959 by Pfeifer and Tappeiner [2] as angioendotheliomatosis proliferans systemica. The origin of the mononuclear cells is controversial. Angiotropic lymphoma is now considered to be of lymphocytic origin and a form of non-Hodgkin’s lymphoma [3, 4]. B cells predominate in this form of malignant lymphoma. The reason that these cells proliferate only within the vascular lumen is unknown. The prognosis is generally poor, with a mean survival of 13 months [1]. The brain and skin are the most commonly affected sites, followed by the adrenals. Lung involvement is not common, but when it occurs it can cause fatal respiratory failure. A few reports of lung involvement in angiotropic lymphoma have emphasized the clinical course and pathology of the disease but not the imaging manifestations [5-7].

In our case, high-resolution CT at a relatively early stage of disease showed multifocal ground-glass opacities with weblike intralobular interstitial thickening and possible narrowing of pulmonary veins. CT scanning at a late stage of the disease revealed widespread ground-glass opacities, suggesting extensive progression of the disease. Although the ground-glass opacities resemble...
hemorrhage, edema, or infarction associated with pulmonary emboli. Alveolar spaces were almost intact at autopsy. Therefore, we consider the ground-glass opacities to be due to interstitial thickening rather than alveolar space filling. The pathologic findings in this case were similar to those in previous reports [5, 6]. We assume that ground-glass opacities with weblike intralobular interstitial thickening are characteristic CT findings.

CT scanning also revealed possible narrowing of the pulmonary veins throughout the lungs. This unique finding probably indicates decreased pulmonary perfusion resulting from intravascular opacification of the lymphoma cells at the pulmonary capillaries. Narrowing of pulmonary veins can be noted in other pulmonary diseases such as cor pul-
monale with pulmonary hypertension and venoocclusive disease. However, diffuse narrowing of the pulmonary veins may be a feature of this disease.

Diagnosing lung involvement in angiotropic lymphoma is difficult because the disease is seldom associated with lymphadenopathy, and its clinical course is often rapidly progressive. However, the combination of ground-glass opacities and narrowing of the pulmonary veins suggests the possibility of lung involvement in angiotropic lymphoma.

References

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