A Case of Pulmonary MALT Lymphoma Arising from Lymphocytic Interstitial Pneumonitis

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Pulmonary mucosa-associated lymphoid tissue-derived (MALT) lymphoma is a rare disease. This disorder is considered to be a model of antigen-driven lymphoma, which is driven either by autoantigens or by chronic inflammatory conditions. Low-grade B-cell MALT lymphoma may develop from a nonneoplastic pulmonary lymphoproliferative disorder, such as lymphocytic interstitial pneumonitis (LIP). A recent estimate predicts that less than 5% of LIP patients acquire malignant, low-grade, B-cell lymphoma. In Korea, there has been no previous report of malignant low-grade, B-cell lymphoma, acquired from LIP. Here, we present the case of a patient with LIP that developed into pulmonary MALT lymphoma, six years after diagnosis.

Key Words: Lymphoma; Lymphoproliferative Disorders; Lung Diseases, Interstitial; Lymphoid Tissue

Introduction

There are various lymphoproliferative disorders which affect the lung parenchyme. Among these, primary pulmonary lymphomas (PPLs) represent only 3 ～ 4% of extranodal non-Hodgkin’s lymphoma (NHL). Pathologically, Low grade B-cell NHL accounts for 58 ～ 72% of cases of PPL. Mucosa-associated lymphoid tissue-derived (MALT) lymphoma is a low grade B-cell extranodal lymphoma characterized by a proliferation of clonal marginal zone lymphocytes that arise from bronchial-associated lymphoid tissue. Stimulation of bronchus-associated lymphoid tissue by antigens, smoking, inflammatory disorders, or autoimmune diseases is thought to be the initial event leading to the development of MALT lymphoma. In rare cases, a nonneoplastic pulmonary lymphoproliferative disorder (p-LPD), such as lymphocytic interstitial pneumonitis (LIP) can develop into malignant, low-grade, B-cell lymphoma. A recent and realistic estimate is closer to 5% of patients with LIP acquire malignant, low-grade, B-cell lymphoma. In Korea, there has been no previous report of pulmonary MALT lymphoma acquired from LIP. We describe here, a rare case of pulmonary MALT lymphoma that arose from a LIP.

Case Report

A 60-year-old woman was admitted to our hospital with cough and dyspnea. At the time of admission, her medical history included medically controlled essential hypertension, diabetes mellitus and old cerebral infarction. She was a smoker of 10 pack-years. Physical examination revealed that she had no skin lesions or neurological abnormalities. There was no lymphadenopathy in the cervical, axillary, or inguinal regions. On auscul-
tation of the chest, a coarse breathing sound with crackles in both lower lung fields was heard. Chest radiographs and computed tomography (CT) scans showed diffuse bilateral ground-glass opacity with intralobular septal thickenings and consolidative lesions (Figure 1A ∼C). Laboratory investigations revealed a total leukocyte count of 7,200 cells/mm³, comprising 40.8% of neutrophils, 49.7% lymphocytes, 7.2% monocytes, 1% basophils, and 1.3% eosinophils. The level of C-reactive protein was slightly elevated to 2.39 mg/dL. The serum lactate dehydrogenase level had increased to 543 U/L (normal, 110 ∼ 230 U/L). Arterial blood gas analysis in room air showed a PaO₂ of 56.5 mm Hg, PaCO₂ 39.3 mm Hg, SaO₂ 90.5% and pH 7.41. Autoantibody tests were negative. Cellular analysis of the bronchoalveolar lavage fluid (BALF) revealed a composition of 25% lymphocytes, 3% eosinophils, 0% neutrophil, and 72% macrophages. Histological examination of video-associated thoracoscopic lung biopsy specimens from the left upper lobe revealed diffuse severe lymphoid cell infiltration with some plasma cells in the interstitium and alveolar walls (Figure 2A). Immunohistochemical examination revealed lymphoid cells evenly admixed with B lymphocytes and T lymphocytes, which were immunoreactive for CD79a (Figure 2B) and CD3 (Figure 2C), respectively. These findings were consistent with the diagnosis of LIP with follicular bronchiolitis components.

Our patient was treated with prednisone 1 mg/kg per day with gradual tapering for 8 months. She showed progressive clinical and radiographic improvement (Figure 1D).

Five years later, at the age of 65 years, our patient started re-experiencing dyspnea and cough symptoms. A chest CT scan revealed diffuse bilateral ground-glass opacity with fine reticulation, traction bronchiectasis and consolidative lesions (Figure 3A, B). Pulmonary