Lymphocytic Interstitial Pneumonitis Presenting as Recurrent Pneumothoraces*

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A 34-year-old woman seropositive for the human immunodeficiency virus presented with recurrent, bilateral pneumothoraces. She also had bilateral interstitial and alveolar infiltrates, and histologic examination was consistent with lymphocytic interstitial pneumonitis. To our knowledge, this is the first documented case of lymphocytic interstitial pneumonitis presenting with recurrent pneumothoraces.

(Chest 1991; 100:1733-35)

Lymphocytic interstitial pneumonitis (LIP) is a rare pulmonary lymphoproliferative disease affecting primarily the alveolar septae and peribronchial tissue. This reaction is composed of a mixture of mature lymphocytes and plasma cells, and is differentiated from lymphoma by the polymorphic nature of the infiltrate and the absence of lymph node involvement. The usual clinical presentation is one of cough, dyspnea, and weight loss. Other less common symptoms include low-grade fever, recurrent pneumonitis, pleuritic chest pain, and fatigue. This report describes a patient with histologically confirmed LIP who presented with recurrent bilateral pneumothoraces.

CASE REPORT

A 34-year-old woman presented to the emergency department with the sudden onset of dyspnea. A chest roentgenogram (CXR) revealed a large right pneumothorax and a chest tube was inserted (Fig 1). A repeated CXR showed reexpansion of the right lung and bilateral interstitial and alveolar infiltrates without mediastinal or hilar adenopathy. Sputum studies for bacteria, mycobacteria, fungi, and Legionella species were negative and the patient refused bronchoscopy. After resolution of her pneumothorax, she was discharged from the hospital on a 30-day regimen of oral penicillin.

The patient returned 45 days later complaining of a persistent cough productive of mucoid sputum, occasional fever, and night sweats. A CXR revealed bilateral alveolar infiltrates and a recurrent 20 percent right pneumothorax. She was admitted to the hospital, and a chest tube was inserted with reexpansion of the lung. Physical examination at that time was pertinent for a very thin woman with bitemporal wasting, bibasilar crackles, dullness at the left base, and a 1/6 systolic murmur at the cardiac apex without radiation. Laboratory results at this time were significant for normocytic anemia with a hemoglobin level of 11.0 g/dl, a negative human immunodeficiency virus (HIV) serologic test, negative tuberculin skin test with positive anergy panel, and an arterial oxygen tension of 71 mm Hg on room air. Immunoglobulin studies revealed polyclonal gammopathy with an elevated serum IgA of 382 WHO mg/dl (range, 63 to 277 mg/dl), elevated serum IgG of 3,000 WHO mg/dl (range, 723 to 1,685 mg/dl), and normal IgM level.

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Lymphocytic lung expansion was difficult to maintain due to a persistent bronchopleural fistula. An additional chest tube was placed, and tetracycline pleuritis was attempted without success. She then underwent thoracotomy, where the left lung was noted to have multiple pleural adhesions. The apical segment of the left upper lobe contained a subpleural bleb with a persistent air leak that was repaired. The left lower lobe was atelectatic with diffuse induration. Biopsy results from the upper and lower lobes were again consistent with LIP. Immunohistochemistry studies of lung tissue revealed a predominance of T lymphocytes (with slightly more suppressor T lymphocytes than helper T lymphocytes), and rare B lymphocytes.

Postoperatively, an anteromedial pneumothorax persisted on the left side despite continued thoracotomy tube drainage. The indwelling chest tube was repositioned and an anterior chest tube was placed with approximately 90% reexpansion of the left lung. Prednisone therapy, 15 mg by mouth every 8 h, was begun. The persistent left pneumothorax gradually resolved, and both chest tubes were removed after two weeks without recurrent pneumothorax. She was discharged home on a regimen of 15 mg of prednisone three times a day. Physical examination at the time of hospital discharge revealed persistent fine bibasilar crackles and signs of consolidation in the left posterior lung base.

At follow-up one month after beginning corticosteroid therapy, the patient reported symptomatic improvement and had gained 0.9 kg. She had only mild crackles bilaterally. Her CXR showed postoperative changes and marked clearing of the alveolar infiltrates with persistent but improved interstitial infiltrates, predominantly in the lower lobes.

**DISCUSSION**

LIP is a rare lymphoproliferative disease of the lungs, first described by Liebow and Carrington in 1966, and more recently recognized in association with the acquired immune deficiency syndrome (AIDS). The cause is unclear. Some authors postulate a systemic immune response to an as yet undefined antigen, as LIP often occurs concurrently with the systemic disease Sjögren's syndrome. Others speculate that LIP is a local hypersensitivity reaction to an inhaled stimulus because it may present in the absence of systemic manifestations. In conjunction with AIDS, LIP has been postulated to be a humoral mediated reaction to the HIV itself or to the Epstein-Barr virus, which is frequently present in the lungs in association with HIV.

LIP occurs predominantly in women with a 2 to 1 female to male ratio. The age of onset ranges from 6 months to 77 years, but affected patients most often present between ages 30 and 60 years. Presenting symptoms are usually dyspnea on exertion, cough, and occasionally weight loss. The classic roentgenographic findings are bilateral interstitial infiltrates of the lower lobes. Alveolar and reticulonodular infiltrative patterns are also seen, and in end-stage disease, a honey-combing pattern may predominate. Mediastinal or hilar adenopathy usually implies the presence of frank lymphoma. LIP causes a restrictive lung disease pattern on pulmonary function testing with a decreased diffusing capacity. The disease is frequently severe enough to produce exercise-induced hypoxemia.

LIP has been observed in association with Sjögren's syndrome with one third of all cases of LIP being found in conjunction with this disease. It also occurs frequently in pediatric AIDS patients. Three quarters of all children with AIDS who are younger than 16 years old have LIP. In adult AIDS patients, the frequency of LIP appears to be much lower. A dysgammaglobulinemia syndrome is frequently linked to LIP. LIP has also been described in association with thyroiditis, chronic active hepatitis, pernicious anemia, autoerythrocytic purpura, and diffuse lymphoid hyperplasia.

Treatment with corticosteroids has been tried, but the efficacy is variable, and spontaneous regression of the disease does occur. A few anecdotal reports of the use of immunosuppressive agents also exist, but responses are difficult to evaluate. In LIP associated with hypogammaglobulinemia, gamma globulin therapy has been reported to be of some efficacy.

Our patient's presentation with bilateral recurrent pneumothoraces secondary to LIP is unique and not previously described. The pathologic changes leading to pneumothorax in our patient are not entirely clear, as parenchymal necrosis is not ordinarily a component of this disease. However, any severe inflammatory process of the lungs presents a potential for pneumothorax, especially when subpleural lung parenchyma is involved. Given the severity and extensive nature of this patient's disease, we speculate that extensive parenchymal involvement with destruction of subpleural pulmonary architecture and visceral pleural disruption were probably underlying factors predisposing her to recurrent pneumothoraces. Her case is typical of LIP in several other aspects. She is female, in the appropriate age group, and has a clinically silent dysgammaglobulinemia.

LIP remains a rare entity. However, as the incidence of AIDS increases, additional patients with unexplained recurrent pneumothoraces may be encountered. While not the most common etiologic entity, LIP should be a diagnostic consideration in these patients.

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Palliation of Left Main Bronchus Compression Due to Malignant Tumor by Intubation via a Tracheostomy Tube*

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Intubation of the left main bronchus via a tracheostomy tube was performed in a patient with local recurrence of lung cancer associated with invasion and obstruction of the left main bronchus after right sleeve pneumonectomy. The result was satisfactory not only for preventing asphyxia, but also for maintaining the patency of the airway after extubation of the endotracheal tube. (Chest 1991; 100:1735-37)

Various methods have been reported for the management of major airway obstruction caused by malignancy, including radiation or laser treatment, coring out under

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FIGURE 1. View of the carina with a flexible fiberoptic bronchoscope. The lesion is extrinsic to and narrows the left main bronchus; the fiberoptic scope could not fit into it.

FIGURE 2. Endotracheal tube (inner diameter, 6.5 mm; outer diameter, 8.5 mm) inserted into the left main bronchus via a tracheostomy tube (inner diameter, 10.0 mm).