Primary Pulmonary Lymphoma With Pleural Involvement as the First Sign of Acquired Immunodeficiency Syndrome

To the editor: Patients infected with human immunodeficiency virus (HIV) have a greater tendency to develop non-Hodgkin’s lymphoma (NHL), with extranodal lesions in more than 90% of the cases. While autopsy series show pleural-pulmonary involvement in 70% of patients with associated systemic NHL, primary pulmonary lymphoma (PPL) is not common among them. Only about 30 cases have been reported (MEDLINE, 1980-2002; key words: pulmonary lymphoma, HIV, AIDS). The case we report is exceptional in two ways: PPL was the first manifestation of acquired immunodeficiency syndrome (AIDS) and lymphomatous involvement of the pleura was detected.

A 60-year-old woman with no known relevant medical history presented with cough, expectoration, shortness of breath with effort, anorexia, and weight loss of 7 kg in the course of 1 month. Exploration showed body temperature to be 38°C, absence of oral candidiasis, slightly enlarged lateral cervical and axillary lymph nodes, and evidence of right pleural effusion. A chest x-ray showed increased opacities and pleural effusion in the right inferior lobe. These findings were confirmed by the computed tomography (CT) scan of the chest in which the pulmonary lesion was seen to be a heterogeneous cavitated mass without adrenal gland enlargement (Figure). Relevant laboratory findings were leukocyte count 3.6×10^9/L; hemoglobin 9.8 g/dL; mean corpuscular volume 81 fl; platelets 380×10^9/L; lactate dehydrogenase 729 U/L; and proteins 6.7 g/dL. The pleural fluid red cell count was 0.56×10^9/L; leukocytes 0.05×10^9/L (90% lymphocytes); glucose 95 mg/dL; proteins 5.18 mg/dL; lactate dehydrogenase 1016 U/L; adenosine deaminase 32.5 U/L; and pH 7.44. Bacterial cultures were negative and no atypical cells were detected. No endobronchial lesions were evident by bronchoscopy, and microbiology assays and cytology of bronchial aspirates were negative. The pleural needle biopsy indicated nonspecific pleuritis. The patient remained feverish throughout 3 weeks of treatment with amoxicillin-clavulanic acid. A thoracotomy was performed and the right inferior lobe was removed. The histologic diagnosis after examination of the lung and parietal pleural tissue was highly malignant type B NHL. A bone marrow sample, an abdominal CT scan, and a cerebrospinal fluid sample were normal. A test for anti-HIV antibodies was positive, the CD4 lymphocyte count was 0.10×10^9/L, and the HIV load was 40 000 copies/mL. The patient did not report drug dependence, although she did mention having
multiple heterosexual partners. She accepted a highly active antiretroviral treatment, but not chemotherapy. Eighteen months after diagnosis the tumors had spread, she was readmitted and died.

A firm diagnosis of PPL requires the absence of mediastinal and hilar lymph node involvement, with no thoracic spread in the 3 months after the preliminary diagnosis. PPL is a rare disease that accounts for fewer than 1% of NHL cases. Most cases of PPL are B cell tumors with a low level of malignancy (bronchus-associated lymphoid tissue or mucosa-associated lymphoid tissue lymphomas). However, in immunodeficient patients, highly malignant, large B cell histological forms are more common. In a group of 12 patients with AIDS-associated PPL, HIV diagnosis occurred from 1 to 8 years prior to the diagnosis of PPL. Only 3 of the patients did not have AIDS when they developed PPL, and all were severely immunosuppressed, with CD4 lymphocyte counts of less than 0.05×10⁹/L. The radiological manifestation of PPL consists of unilateral and bilateral nodes or of a subpleural pulmonary infiltrate or mass that may cavitate. Although pleural effusions have been described in 15% of immunocompetent patients with PPL, such effusion was detected in only 1 of the HIV positive patients in the previously mentioned study, and this patient was probably the same one referred to by the same authors in another series in which it was seen that the pleural effusion occurred during the postoperative phase following pulmonary biopsy. Lymphomatous involvement of the pleura was not observed during the autopsy. In our patient, the pleura might have become involved because it was adjacent. The diagnosis of PPL requires transthoracic needle biopsy of the lung, thoracoscopy, or thoracotomy. The mean survival rate in the aforementioned group of patients was 4 months (range <1 to 17 months), and patients died from disease progression or from opportunistic infections. Treatment must be specific to every patient but includes the combination of antiretroviral drugs, surgery, and chemotherapy.

J.M. Porcel Pérez and M. Rubio Caballero Servicio de Medicina Interna, Hospital Universitario Arnau de Vilanova, Lleida, Spain.