Langerhans-Cell Histiocytosis of 28-Year Evolution

To the Editor: Pulmonary Langerhans-cell histiocytosis (PLCH) belongs to a spectrum of diseases characterized by monoclonal proliferation and invasion of various organs by Langerhans cells. Little is known of the natural history of PLCH and the prognosis is variable and unpredictable in adults, in whom survival has been estimated to be 12.5 years.[1] We report the case of a patient with PLCH of 28-year evolution.

A 57-year-old man was referred in 1975 with a 2-year history of breathlessness related to exercise. He was a smoker who worked installing artificial stone flooring. Reticulonodular infiltrates were observed on a chest x-ray and lung function tests revealed a mild restrictive pattern, with decreased carbon monoxide diffusing capacity (DLCO) and increased lung volumes. A lung biopsy procedure was performed and the histologic diagnosis was diffuse alveolar fibrosis. After a year of treatment with oral corticosteroids, the patient was asymptomatic, although he continued to smoke. He had occasional, irregular check ups at work, sometimes presenting productive cough and wheezing. Obstructive-type functional alterations were noted as well as the same findings of increased lung volumes and decreased DLCO. Reticulonodular images continued to be present in radiographs. A high resolution computed tomography (HRCT) scan of the chest had recently revealed evidence of diffuse central emphysema mainly in the upper lobes (Figure). The lack of consistency between that radiographic finding and the 28-year-old histologic diagnosis led to reconsideration of the diagnosis, and re-examination of the specimen obtained earlier led to a diagnosis of PLCH.

PLCH is a rare disease—accounting for 5% of cases of diffuse interstitial lung diseases[2]—and its etiology is unknown. Lung function tests at disease onset may be normal or show a pattern of mild restrictive, obstructive, or mixed alterations. DLCO is diminished in 60% to 90% of cases. As the disease progresses, findings will suggest either a restrictive pattern, if fibrosis predominates, or an obstructive one of varying severity, if cysts predominate. Chest radiographs usually give positive findings, the most common image being a micronodular or reticulonodular pattern in the middle and upper lobes. HRCT is superior to simple radiography for diagnosis, and a finding of confluent irregular cystic spaces with peribronchial nodular opacities in the middle and upper lobes is highly indicative of PLCH.[3] Having the patient stop smoking is the most important therapeutic step to take. The effectiveness of corticosteroids is debated, although they continue to be the most widely prescribed treatment in patients who are symptomatic or whose disease is progressing. Disease course is variable and unpredictable, with reports of both asymptomatic patients and patients in whom progression leads rapidly to respiratory failure and death.[4] There are reports of cases that remitted or stablilized, followed by recurrence after periods that have varied from a few months to up to 25 years.[5] Diminished DLCO, low forced expiratory volume in the first second, and increased residual lung volume found at disease onset have been considered predictors of poor prognosis. Likewise, the prognosis for smokers is considered poor.

The patient we have described has been diagnosed as having PLCH. The long history of altered lung function test findings and the images provided by HRCT mean that it is unlikely that cure took place, indicating that the alterations found 28 years after initial presentation were the result of pre-existing lesions caused by the disease, on which smoking-related emphysema can be said to have been superimposed. The case is unusual for its long evolution in spite of the presence of risk factors suggesting poor prognosis.

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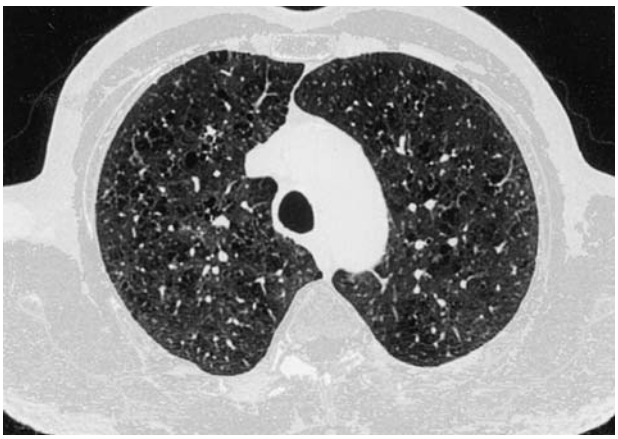

Figure. High resolution computed tomography scan of the chest in which cystic spaces with poorly defined borders can be seen in both upper lobes.