Active pulmonary tuberculosis in a patient with Langerhans’ cell histiocytosis

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Langerhans’ cell histiocytosis (LCH) or pulmonary histiocytosis is an uncommon parenchymal lung disease of unknown origin. The lesions of LCH contain single or multifocal proliferation of histiocytes similar in phenotype to dendritic Langerhans’ cell. The natural history is extremely variable. The disorder ranges in clinical severity from a solitary eosinophilic granuloma of bone to a generalized disease with multiple organ involvement. Rarely, LCH has been reported to be associated with some disorders such as pulmonary tuberculosis. Here, we report a case of a 21-year-old male patient with LCH and active pulmonary tuberculosis.

A 21-year-old male patient was hospitalized due to complaints of progressive dyspnea in the last 6 months, occasional cough and night sweats. There was nothing marked in the medical history of the patient who was working in tourism industry. He had a 9 years smoking history. Physical examination of respiratory system and other systems were normal. Routine hematological studies were normal; there was no peripheral eosinophilia, the erythrocyte sedimentation rate was 35 mm/h. Other hematological and biochemical data on admission were within normal ranges. Postero-anterior (PA) chest x-ray revealed extended heterogeneous reticulonodular infiltration in both lung fields. Thorax high resolution computed tomography (HRCT) revealed a reticulonodular infiltration involving all lung areas in which there were thin walled cavitary lesions among those the biggest one was 10 mm in diameter. Nonspecific culture of sputum revealed normal oral flora. Tuberculin skin test was negative. Acid-fast bacilli (AFB) was not detected in the sputum, in gastric lavage, and in bronchial washing fluid under microscopic examination. A fiber optic bronchoscopy was performed. There was no endobronchial pathology. Brochoalveolar lavage fluid examination and cell distribution were nondiagnostic. Transbronchial biopsy specimens showed normal lung mucosa and parenchyma. Pulmonary function tests (PFT) revealed restrictive disease. The PFT presented the following values: Vital capacity (VC), 3.45 L (62% pred); forced vital capacity (FVC), 2.85 L (56% pred); FEV1, 2.79 (78% pred); and peak expiratory flow rate, 6.25 L/sec (71% pred). Diffusion capacity was lowered (4.9 mmol/min/kPa, 62% pred).

The patient underwent a video assist thoracic surgery for definite diagnosis. Histopathological examination of the specimen of biopsy revealed many eosinophils, histiocytes and lymphocytes all of which showed centrilobular distribution pattern consisting of dense chronic inflammation, edema, and fibrosis. Muscular hyperplasia and dilatation were detected. Immunohistochemical study of S-100 protein of biopsy specimen showed S-100 protein positive histiocytes in granulomatous lesions. In addition, moderate interstitial fibrosis was seen with trichrome staining. Histological findings were compatible with LCH. There was no clinical or laboratory finding compatible with LCH in other organs. Abdominal ultrasound was normal. During laboratory evaluation Mycobacterium tuberculosis was isolated on culture media. The final diagnosis was LCH and pulmonary tuberculosis. Antituberculosis treatment with isoniazid, rifampicin, pyrazinamide and ethambutol was started. Corticosteroids were not used for the treatment. A repeat thorax HRCT examination after 6 months from the beginning of the treatment revealed marked improvement in nodular lesions, and thin walled cystic lesions in the upper lung fields. Marked regression was detected for micro nodular opacities in both lung fields. Our patient was diagnosed with pulmonary LCH proven through lung biopsy and AFB culture positive pulmonary tuberculosis. Antituberculosis treatment resulted in an objective radiological improvement and AFB culture negativity. The clinical course of LCH ranges from spontaneous resolution to a chronic and sometimes lethal disease. The etiology of LCH remains obscure, but the vast
majority (up to 97%) of patients are current smokers or ex-smokers. Although, spontaneous improvement in patients with LCH has been described, the role of smoking cessation in the natural history remains controversial. After diagnosis, our patient stopped smoking. In pulmonary LCH, chest x-ray usually reveals a diffuse reticulonodular pattern with micronodules measuring 2-5 mm in diameter and pulmonary cysts that predominate in the upper lobes. However, in occasional cases chest x-ray can be normal and CT scan of the lung may disclose focal or diffuse abnormalities. In pulmonary LCH cases, it was observed that nodular opacities, thick-walled cysts, and ground-glass opacities underwent regression. Thin-walled cysts, linear opacities, and emphysematous lesions remained unchanged or progressed. In active pulmonary tuberculosis cases, CT scan reveals combination finding of centrilobular nodule or branching linear structure, bronchial wall thickening. In the present case, HRCT scan revealed a reticulonodular infiltration involving all lung areas in which there were thin-walled cavitary lesions. Due to the high incidence of spontaneous remission in untreated patients, the efficiency of different immunosuppressive therapeutic strategies for pulmonary LCH remains unknown. Since severe impairment of pulmonary function at the time of diagnosis has negative prognostic value in patients with LCH an early intervention may be beneficial in terms of survival. In our patient, we have shown substantial clinical and radiological improvement after antituberculosis treatment, and cessation of smoking without corticosteroid treatment. It was not revealed if the improvement were due to antituberculosis treatment or cessation of smoking. Reviewing the literature, we identified only one published LCH case with pulmonary tuberculosis.

In summary, this case demonstrated that LCH could be associated with active pulmonary tuberculosis. Thus, it should be kept in mind that LCH and pulmonary tuberculosis might exist as concomitant disorders in the same patient particularly in areas where prevalence of tuberculosis is high.

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