CASE REPORT

Idiopathic Bronchiolocentric Interstitial Pneumonia: A New Idiopathic Interstitial Pneumonia

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Idiopathic interstitial pneumonias represent a diverse group of lung diseases with diffuse effects on the lung parenchyma. In 2002, the American Thoracic Society/European Respiratory Society consensus classification unified the descriptions of the different entities encompassed by idiopathic interstitial pneumonias. Despite this broad consensus there are still some entities without a clear definition and others, such as idiopathic bronchiolocentric interstitial pneumonia, that were only later described. We present the case and outcome of a woman diagnosed with idiopathic bronchiolocentric interstitial pneumonia by lung biopsy.

Key words: Idiopathic bronchiolocentric interstitial pneumonia. Idiopathic interstitial pneumonia. Interstitial lung disease.

Introduction

Idiopathic interstitial pneumonias represent a heterogeneous group of diseases with diffuse effects on the lung parenchyma, characterized by varying degrees of inflammation and fibrosis. They are classified as different diseases that present a number of specific characteristics, including clinical presentation, radiological pattern, histologic pattern, response to treatment, and prognosis.

In 2002, a joint statement of the American Thoracic Society (ATS) and the European Respiratory Society (ERS) unified the classification of the different diseases represented by idiopathic interstitial pneumonia by means of a set of clinical, radiological, and histologic descriptions. Despite this broad consensus, there are still some entities without a clear definition, such as nonspecific interstitial pneumonia, and even some interstitial pneumonias that do not fit any of the described patterns. Furthermore, there are other entities that are not included in this consensus as they were only described later. This is the case of idiopathic bronchiolocentric interstitial pneumonia.

We present the case of a 37-year-old woman with idiopathic interstitial pneumonia diagnosed by video-assisted thoracoscopy.

Case Description

A 37-year-old woman was admitted to our department with breathlessness. She was an ex-smoker of 5 pack-years and presented no other relevant history of disease. The patient worked as a homemaker and occasionally in agriculture (cherry trees and almond trees) but was not in contact with insecticides. She had no relevant family history and was not taking any regular medication. The patient stated that she had been in contact with dogs, cats, and pigeons. A month before admission, she had begun to experience dyspnea that progressed to dyspnea with minimal effort. The patient presented no other symptoms.
Discussion

Idiopathic interstitial pneumonias are inflammatory diseases of the lung parenchyma that are differentiated by the histologic pattern, the distribution of the disease in the lung, which lobe is primarily affected, the maturity of the interstitial fibrosis, and the presence of certain characteristic histologic findings, such as granulomas or the presence of macrophages in the alveoli. Among the idiopathic interstitial pneumonias described in the ATS/ERS consensus statement, only bronchiolitis associated with interstitial lung disease has a predominately bronchiolocentric distribution. Several authors have recently described a new interstitial pneumonia with predominately bronchiolocentric involvement and no granulomas.2-3 Yousem and Dacic2 described the characteristics of 10 patients whose histologic pattern was characterized by a chronic inflammatory cell infiltrate of centrolobular and bronchiolocentric origin. In those patients, the small airways presented a lymphocytic and plasma cell infiltrate between the peribronchial alveolar septa and in the alveolar interstitial space, along with fibrosis and/or peribronchiolar metaplasia. Those authors found a higher proportion of women in their case series and radiography revealed an interstitial infiltrate predominantly affecting the lower lung fields. The physical examinations revealed a restrictive pattern with diffusion abnormalities. All the patients were treated with corticosteroids and/or immunosuppressants and prognosis was poor in most cases. Churg et al3 described a series of 12 patients, also predominately women and with a poor prognosis. The histologic pattern was similar to that described by Yousem and Dacic2 but with more severe peribronchial fibrosis and greater thickening of the bronchial muscle. Fukuko et al4 described a histologic pattern characterized essentially by the presence of peribronchiolar metaplasia. Several of the patients included in that series presented rheumatological diseases that may have been the cause of the lung involvement. The centrolobular fibrosis described by de Carvalho et al5 in their series of 12 patients was characterized by the presence...
of bronchial necrosis and bronchiolocentric fibrosis. Foreign bodies were present in 42% of the patients in that series and the authors indicated that the histologic pattern was secondary to gastric aspiration. It is possible that the different histologic patterns described by these authors correspond to different stages of the same entity. In this case, the bronchiolar metaplasia described by Fukuoka et al would be the mildest stage, followed by the idiopathic bronchiolocentric pneumonia described by Yousem and Dacic. The most severe level of fibrosis would be the pattern described by Churg et al. In our patient, the radiological, functional, and histologic patterns and the course of the disease were similar to those of the cases of bronchiolocentric interstitial pneumonia described by Yousem and Dacic.

Assessment of idiopathic interstitial pneumonias should rule out diseases that may be a potential cause of these patterns of inflammation and pulmonary fibrosis, particularly, autoimmune diseases, allergic and drug reactions, and pneumonitis. In our case, in addition to ruling out the presence of autoimmune diseases, possible drug side effects were excluded on the basis that the patient was not undergoing any pharmacological treatment. In the case of idiopathic bronchiolocentric interstitial pneumonia the differential diagnosis should particularly include hypersensitivity pneumonitis and nonspecific interstitial pneumonia. Our patient was in contact with pigeons but precipitins against pigeon antigen were negative and the patient did not improve on avoiding contact. Furthermore, the histologic study did not reveal the presence of the granulomas characteristic of hypersensitivity pneumonitis (necessary for diagnosis in the opinion of some authors).

Nonspecific interstitial pneumonia is characterized by good prognosis and response to treatment and the histologic pattern is characterized by the lack of bronchiolocentric involvement. Our patient did not improve following a year of treatment and the histologic pattern was characterized by predominately bronchiolocentric involvement.

After evaluating the radiological, functional, and histologic patterns and the course of the disease, we conclude that our patient presented idiopathic bronchiolocentric interstitial pneumonia.

REFERENCES