European Respiratory Society
Annual Congress 2012

Abstract Number: 3077

Publication Number: P3626

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Interstitial lung disease **Keyword 2:** No keyword **Keyword 3:** No keyword

Title: Airway-centered interstitial fibrosis – Two case reports

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Body: Airway-centered interstitial fibrosis (ACIF) is a rare interstitial lung disease (ILD) of unknown cause characterized by chronic cough and progressive dyspnea and history of inhaled exposure. There is progressive peribronchiolar distribution of interstitial inflammation and fibrosis with bronchiolar metaplasia. The majority of patients are non-responsive to corticotherapy and prognosis is poor. We describe two female patients, 65 and 66 years, presenting with chronic dry cough and progressive dyspnea. They were non-smokers, farmers and with a history of inhaled exposure to birds. Pulmonary function tests showed a moderate obstructive pattern in one case and moderately decreased CO diffusion in both patients. Chest radiographs revealed diffuse reticulonodular infiltrates. Chest computed tomography (CT) showed reticular fibrosis and disperse ground glass infiltrates in one case and sparing the upper lobes in the other. Bronchoalveolar lavage showed an increase in lymphocytes and neutrophils in one case. The diagnosis was made by surgical biopsy revealing pericentrilobular lesions and linfoplasmocitary infiltrate compatible with ACIF. Treatment with oral steroids for 12 months in both patients. In one case the disease progressed with worsening symptoms, pulmonary function tests and CT imaging even after combined immunosuppression with Azathioprine. In the other case the patient's symptoms and CT imaging improved. These cases had similar clinical, radiological and pathological features as most of the few cases reported in literature. Both were non-smokers but had been exposed to birds. However, disease progression varied considerably as one patient improved but the other worsened even after combined immunosuppression.