Pulmonary Langerhans Cell Histiocytosis

Abstract:
M Kooblall, S Hanad, E Moloney, SJ Lane
Respiratory Department, AMCR, Tallaght, Dublin 24

Abstract
We report the case of a 57 year old man who presented with increased shortness of breath together with increased pulmonary nodules in his upper lobes over a two year period. His strong smoking history and pattern of distribution makes Langerhans cell Histiocytosis a likely diagnosis that was confirmed on biopsy.

Case Report
This patient has a background of stage D COPD, Obstructive Sleep Apnea and type 2 diabetes. He is a current smoker with 80 pack a year history. He works in an office in a drug rehabilitation centre and is fully independent in his activity of daily living. He initially presented to the outpatient department with exertional dyspnea. He had a CXR done which showed a tiny 4mm nodule in the right lower lobe. Since 2009, he had been on surveillance register. Follow up CT Thorax in June 2011 showed two non specific nodules in the right lower lobe measuring 7mm and 6mm. As per the fleshisser criteria he was booked for a follow up CT Thorax in the following three month time which showed new bilateral upper lobes inflammatory nodules in a peribronchovascular distribution.

Over the intervening time period, there was a deterioration in his functional status as he was getting more breathless on minimal exertion, occasional non productive cough, no chest pain, no fever, no weight loss, no night sweat. His vitals and clinical examination were normal. His FBC, U&F, LFT, ESR, CRP, Tumour markers (PSA, HCG, AFP, CA 19.9, CEA), ACE and mantoux test all came back normal. His quantiferon however came back positive. His PFT done showed: FEV1 -1.25L (33% predicted), FVC - 2.94L (36% predicted), FEV1/FVC - 34%, TLC – 81%, KCO – 95%. His transsthoracic echocardiogram was normal. Bronchoscopy showed a nodule on the vocal cord otherwise no endobronchial lesion found. BAL and transbronchial biopsies all came back normal. After being discussed at our lung cancer MDT, it was agreed to proceed ahead with a right lower lobe wedge biopsy. The microbiopsy came back as follows: airways stellate scars and several nodules present composed of grooved histiocytic cells (CD1a+) and a scattering of eosinophils consistent with Langerhans cell Histiocytosis.

Discussion
Pulmonary Langerhans cell histiocytosis (LCH) in adults (Eosinophilic granuloma, Histiocytosis X) is characterised by monoclonal proliferation and infiltration of organs by Langerhans cells. The lung is the principal site of involvement. The histiocyte society has established a simplified classification ranging from involving single organs to more aggressive multiorgan disease. Its presentation ranges from asymptomatic (25% of cases) to rapidly progressive (non productive cough and dyspnea most common). Immunohistochemical studies are useful in recognizing Langerhans cells, which stain for the S-100 protein, CDla and HLA-DR. However the mere presence of Langerhans cell is not diagnostic of LCH. Ground glass infiltrates may be radiologically indistinguishable from features of hypersensitivity pneumonitis, bronchiolitis obliterans with organising pneumonia or chronic eosinophilic pneumonia. The histological lesion progress from cellular nodules to entirely fibrotic nodules that are often stellate in configuration and may connect with nodules in adjacent lung parenchyma to produce a distinctive honeycomb-like structure with enlargement of air spaces and hyperinflation. In later stages fibrotic nodules may lack Langerhans cells entirely. A number of histologic findings are commonly associated with the lesions of pulmonary LCH. Since the majority of patients are smokers, the findings of respiratory bronchiolitis is not surprising. In some cases respiratory bronchiolitis is sufficiently extensive that the clinical symptoms could be due to interstitial lung disease associated with respiratory bronchiolitis. Filling of the air spaces by pigmented macrophages is common and may cause confusion with desquamative interstitial pneumonia. Emphysematous changes of the underlying lung tissue are also common as is air-space enlargement with fibrosis.

In adults it has been reported to be associated with lymphoma and rarely carcinoma, thereby outlining the importance of early detection. Open or thoracoscopic surgical lung biopsy has the highest diagnostic yield. There is no definitive therapy and therefore discontinuation of smoking should be emphasised as a cornerstone of treatment. Finally its outcome can be variable ranging from spontaneous remission to remission following cessation of smoking to progression to end stage fibrosis.

Correspondence: M Kooblall
Respiratory Department, Tallaght Hospital, Dublin 24
Email: mineshamnch@gmail.com

References