Pulmonary Langerhans Cell Histiocytosis

Evolution of radiologic findings after smoking cessation

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We present the case of a 30 year old male with non productive cough for the last 2 months. No other symptoms were reported. He had been a farmer since the age of 15 and was a current smoker (1 pack of cigarettes for 25 years). His personal medical history was negative and he was on no medication. Physical examination revealed no abnormal findings.

High Resolution Computed Tomography (HRCT) revealed the presence of bilateral and symmetrically distributed innumerable centrilobular nodules and cysts with clearly perceptible walls allowing them to be differentiated from emphysema. The abnormal findings had striking upper lobe predominance, with characteristic sparing of the costophrenic angles. The patient was subjected to bronchoscopy and bronchoalveolar lavage (BAL). The results of BAL were: Macrophages: 78%, Lymphocytes: 18%, Eosinophils: 1%, Neutrophils: 3%, CD1α: 6%. The combination of radiologic and BAL findings secured the diagnosis of Pulmonary Langerhans Cell Histiocytosis (PLCH) obviating the need for tissue confirmation[1]. Smoking cessation was strongly advised. A new HRCT performed 9 months later showed an almost complete resolution of radiographic findings.

It is worth noting that the early "cavitation" of nodules seen in PLCH is due to the bronchocentric localization of inflammation and not to a necrotic process, hence the quotation marks. As the granulomatous inflammation progresses in the peribronchial area, it causes destruction of the bronchiolar wall and dilation of the lumen[2]. The resulting increased contrast in attenuation between the bronchial wall/peribronchial area and the airway lumen gives the impression of early "cavitation". This also explains the radiologic progression of PLCH from nodules to thick wall cysts to thin wall cysts and finally to bizarre shaped cysts.

With this case, we would like to highlight the characteristic HRCT findings of PLCH, the potential diagnostic value of BAL and also the fact that in term of management smoking cessation is of utmost importance[3].

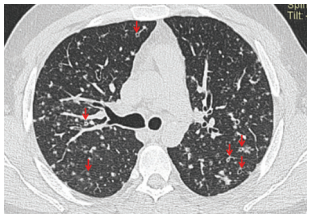

FIGURE 1. Level of Right Upper Lobe bronchus. Bilateral and symmetric distribution of innumerable centrilobular nodules and thick walled cysts (arrows)

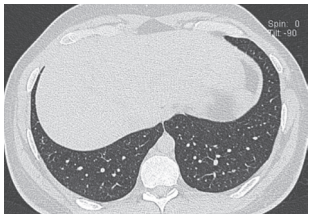

FIGURE 2. Characteristic sparing of the lung bases.

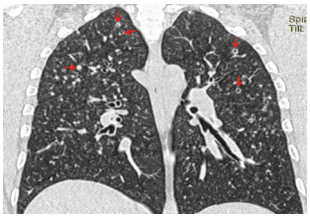

FIGURE 3. Coronal reformation showing centrilobular nodules and thick walled cysts (arrows). The upper/middle zone predominance of the disease with sparing of the lung bases is clearly demonstrated.

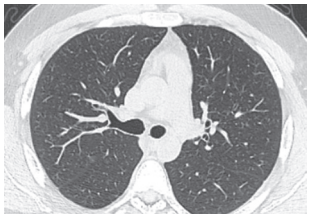

FIGURE 4. HRCT after 9 months in the same axial level as Figure 1. There is an almost complete resolution of abnormal findings.

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