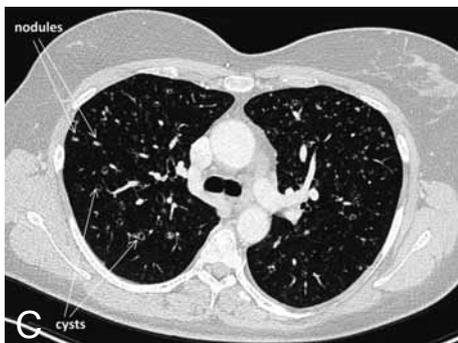
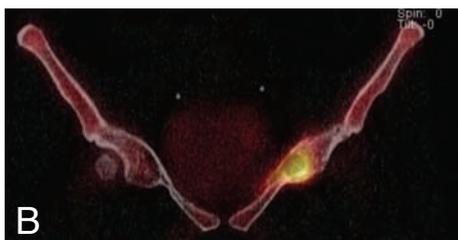


## IMAGES IN CLINICAL RADIOLOGY



### *Pulmonary Langerhans cell histiocytosis and eosinophilic granuloma*

M. Demeter, Y. Vankan, F. De Somer, D. Perdieus<sup>1</sup>

A forty-three-year-old female patient presented at the emergency department for low back pain and left hip pain since four weeks, mostly at night, and disabled walking. Neurological examination and laboratory findings were normal. Computed tomography of the lumbar spine showed degenerative discopathy at level L4-L5 and L5-S1, but revealed no other lesions explaining the patient's acute complaints. Additional plain radiograph of the left hip revealed an oval cyst-like lucent lesion in the left iliopubic ramus with thinning of the adjacent cortical margin (Fig. A). A magnetic resonance imaging (MRI) for further exploration was proposed. The MRI revealed a T1 hypointense lesion surrounded by increased signal intensity on T2 images, consistent to an osteolytic lesion with peri-lesional bone marrow edema, and a concomitant fracture of the iliopubic ramus. SPECT-CT confirmed the presence of a lytic bone lesion in the left iliopubic ramus with high tracer intensity (Fig. B), as well as three smaller osteolytic lesions in both ileum bones without tracer uptake. Considering the possibility of bone metastasis, thoracic and abdominal CT examination was performed to rule out a primary tumor. Both lungs showed multiple small cysts with irregular wall thickness, as well as numerous millimetric pulmonary nodules with groundglass and more dense appearance, both localized in the upper and middle lung zones, with sparing of the lung bases and costophrenic angles (Fig. C). The diagnosis of pulmonary Langerhans cell histiocytosis and eosinophilic granuloma was suggested. An ultrasound-guided percutaneous biopsy of the bone lesion in the left iliopubic ramus was performed. Histological and immunohistochemical examination of the biopt revealed a cluster of eosinophilic granulocysts staining for S100 protein and CD1 antigen, a specific Langerhans cell marker. The diagnosis of 'pulmonary Langerhans cell histiocytosis' and 'eosinophilic granuloma' was confirmed.

#### *Comment*

Pulmonary Langerhans Cell Histiocytosis (PLCH), previously known as histiocytosis X or eosinophilic granuloma of the lung, is a very rare and uncommon interstitial lung disease, affecting young adults between 20 and 40 years old; women sometimes at older age.

In 4% to 20%, eosinophilic granuloma occurs extra-pulmonary, causing a cystic bone lesion, mostly in flat bones, with local pain and pathological fractures as a consequence. The diagnosis is mostly made by chest imaging characteristics and histological confirmation by CD1a positive Langerhans cells in for example bronchoalveolar lavage or surgical lung biopsy. High resolution computed tomography (HRCT) of the lungs shows a typical pattern of centrilobular stellate nodules and, with advancing disease, multiple thin- and thick-walled cysts, both with a mid to upper zone predominance and sparing of the costophrenic angles. Though HRCT

findings are very characteristic, PLCH has to be distinguished from other interstitial lung diseases, like lymphangiomyomatosis (LAM) for example. LAM is also a rare cystic lung disease, with thin-walled cysts, present in the lung bases, and without pulmonary nodules, in the contrary to PLCH. One should not mistake the 'cyst-like' destruction and honeycombing of idiopathic pulmonary fibrosis with confluent cysts in advanced pulmonary histiocytosis since they can have a similar appearance though different distribution pattern. The etiology of PLCH is still unknown, but smoking – also present in our patient's medical history – is thought to be a causal factor of the pulmonary presentation of the disease. Therefore, treatment includes first of all smoking cessation. The use of glucocorticoids is proven to be effective; whereas cytotoxic agents are of limited value. In case of solitary eosinophilic granuloma in bone, local radiotherapy and/or curettage is the best therapeutic option.

#### *Reference*

1. Tazi A.: Adult pulmonary Langerhans'cell histiocytosis. *Eur Respir J*, 2006, 27: 1272-1285.

1. Department of Radiology, Imelda Hospital, Bonheiden, Belgium.