Langerhans cell histiocytosis of the lung with probably skeletal involvement

Abstract

Langerhans cell histiocytosis is characterized by abnormal accumulations of large mononuclear cells forming granulomas in various organs mainly in the lung, bone, or skin. Adult pulmonary Langerhans cell histiocytosis is rare and almost always associated with cigarette smoking; combination with lung and bone simultaneous involvement is even rare. We present a 41 years old male smoker who was diagnosed with pulmonary Langerhans cell histiocytosis by a lung biopsy and manifestations at high resolution computed tomography of the lung. Later technetium-99m methyl diphosphonate bone scintigraphy showed multiple abnormal tracer accumulation of the radiotracer in the skull and a singular focus in a rib.

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Introduction

Pulmonary Lanerghans cell histiocytosis (PLCH) is seen at the age of 20-40 years and involves both male and female. There are computed tomography (CT) findings of nodules and cysts in the lung parenchyma with an upper and mid-zone predominance with cystic lesions becoming more prominent over time [1]. Prognosis of PLCH has indolent course in the majority of patients, in a small minority there may be a regression of lesions and in another small subset progression to end stage fibrosis may occur [1]. A patient with PLCH complicated with bone involvement is presented.

Case report

A 41 years old man, smoker was diagnosed with pulmonary Langerhans' histiocytiosis. The patient was found to have evidence of interstitial lung disease based on chest X-rays CXR and high-resolution CT of the chest about 10 years ago. The patient was transferred from another hospital where he experienced multiple episodes of dyspnea, non-productive cough which caused him unable to perform many daily activities. Pulmonary physical examination re-

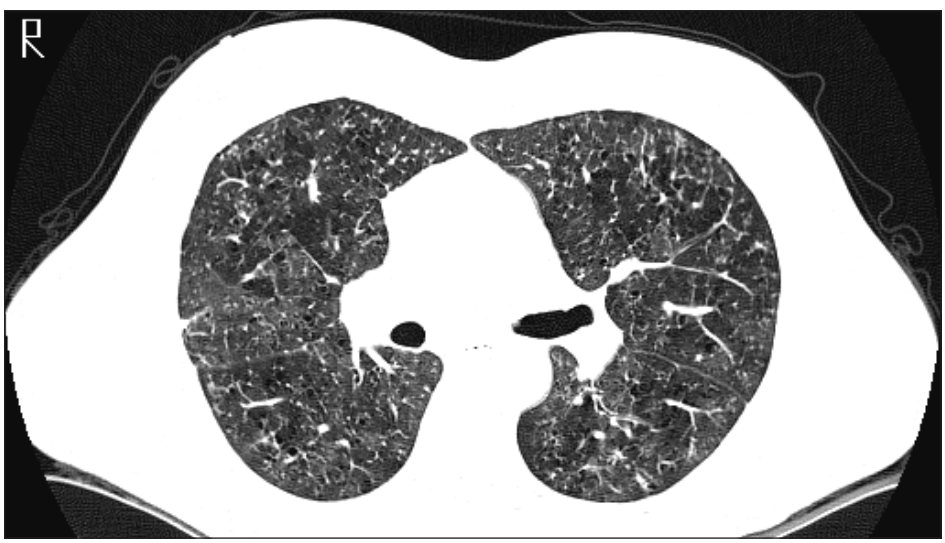

Figure 1. Higher resolution CT shows irregular thin and thick walled cysts of non-uniform shape and upper lobe predominance, small nodular densities and reticular opacities, a mosaic attenuation pattern and scattered areas of ground-glass opacity. These findings are consistent with Langerhans cell histocytosis.

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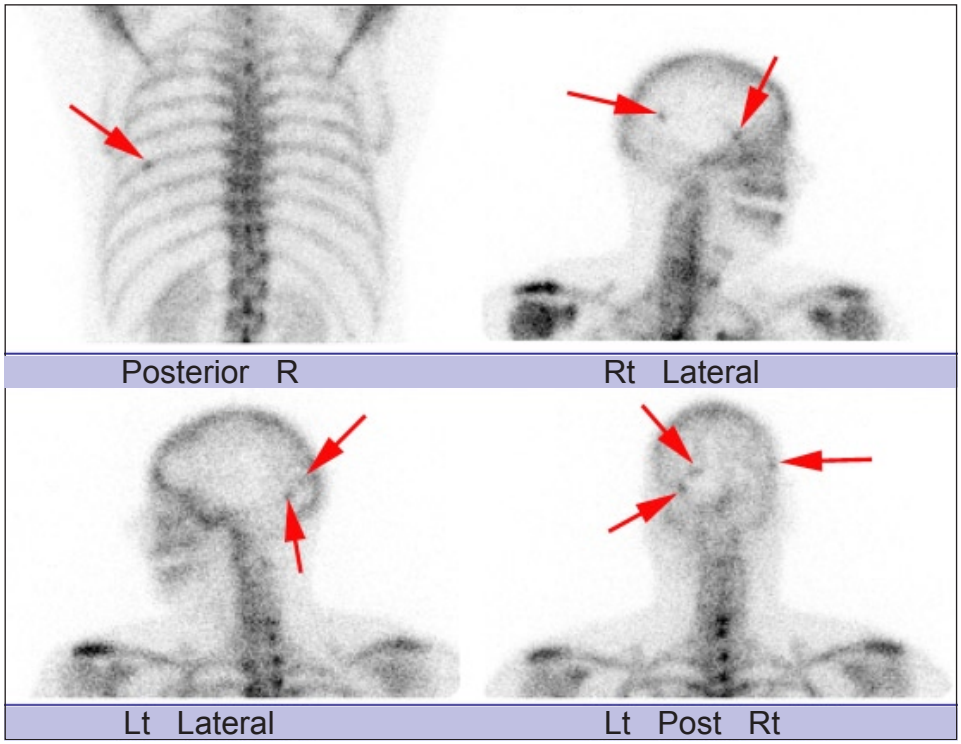

Figure 2. ^{99m}Tc-MDP bone scintigraphies of the head and posterior chest show multiple and mild increased uptake in the skull (arrows) and a mild and focal uptake in the left 8th posterior rib (arrow).

vealed rhonchi, sonorous wheezes on right and left abnormal breathing sounds bilaterally. Multiple bronchoscopic biopsies were non-diagnostic and subsequent evaluation in December 2008 included a video-assisted thoracic surgery with the right middle lung biopsy, on December 04, 2008, that diagnosed Langerhans' cell histocytosis bronchiectasis and patchy lung fibrosis. He continued smoking until 2008 when tissue biopsy was done. Recent high resolution chest CT on December 15, 2009, with 1.3 mm thick images at 1cm interval showed irregular thin and thick walled cysts of non-uniform shape with upper lobe predominance, small nodular densities and reticular opacities, a mosaic attenuation pattern and scattered areas of ground-glass opacity: high-resolution CT findings were consistent with the diagnosis of Langerhans cell histiocytosis (Fig. 1). With complaints of severe bone pain as well as chest muscle pain, he underwent technetium-99m methyl diphosphonate (^{99m}Tc-MDP) total body bone scintigraphy which showed multiple and mild increased uptake in areas of the skull and in the left posterior 8th rib (Fig. 2). He was on Albuterol inhalation every six hours as needed for better breathing.

Discussion

Adult PLCH is a rare disorder of unknown etiology, probably related to immunological factors, that occurs predominantly in young smokers [2]. Five patients with bone involvement have been reported [3]. Involvement of the skull of temporal bone without lung abnormality has also been reported [4], as well as lung and bone (rib) involvement [5]. Pulmonary involvement was the most common location with bony involvement in second place [1]. In our case, adult PLCH with combined skull and rib lesions are presented.

An incidence peak at 20-40 years [1, 2] and an average age 43.5±7.7 years, are common in PLCH. The disease is found 66% in women, and 34% in men [1]. In the majority of cases, the disease follows an indolent course but in a small percentage there may be spontaneous regression with smoking cessation [1, 5-8].

Our patient however continued smoking until 2008 when tissue biopsy was done. Following smoking cessation, lung nodules and cysts gradually disappeared on serial CT scans, with complete clearance of the lesions after 12 months [9].

Skeletal LCH was described by increased uptake on ^{99m}Tc-MDP bone scintigraphy in a pediatric patient [10], and a FDG-avid in proximal femur incidentally found biopsy proven LCH was demonstrated by FD-PET images in a child patient with a small bowel intersusception [11]. However, bone scintigraphy seems to be less sensitive than CT in the detection of this disease [4]. Bone scan does not show significant uptake of the radionuclide due to its lower sensitivity and had been described in comparison with CT of the skull and seems to be less sensitive than radiography in the detection of this lesion [4]. Bone scans are unreliable and easily missing lesions for the detection bone involvement [12-14]. Abnormal histiocytes accumulate in the bone marrow first and do not invade the bone tissue at the initial stage. Thus, bone scintigraphy seems to be less sensitive than bone CT in the detection of LCH bone involvement; total body bone scans, however, may be complimentary with bone CT.

High-resolution (HR) CT has proved a major breakthrough in the diagnosis of PLCH [15], and is now mandatory when this condition in suspected [16-18]. HRCT provides additional details about the parenchymal elementary lesions, such as cavitation of nodules, which is not readily visualization standard radiologaphys [19, 20]. HRCT has led to better appreciation of nodular and cystic radiographic abnormalities characteristic of the disease [21]. Totally 108 cases have been reported in literature [19, 22-33] till today. However, no report is described combination with the lung and bone simultaneous involvement. We are reporting a case of PLCH proved by lung biopsy which was also diagnosed by HRCT and bone scintigraphy and showed multiple abnormal tracer accumulation of the radiotracer in the skull and a rib.

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All authors declare that they have no conflicts of interest

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