Case Report

Diffuse pulmonary uptake of bone-seeking radiotracer in bone scintigraphy of a rare case of pulmonary alveolar microlithiasis

Babak Fallahi, Bahar Moasses Ghafary, Armaghan Fard-Esfahani, Mohammad Eftekhari

Research Center for Nuclear Medicine, Tehran University of Medical Sciences, Tehran, Iran

ABSTRACT

Pulmonary alveolar microlithiasis (PAM) is a rare diffuse pulmonary disease representing microliths formed by deposition of calcium phosphonate in the alveolar airspaces. PAM is often diagnosed incidentally during chest X-ray imaging. Most of them are asymptomatic. We present a 39-year-old man referring for a bone scan due to a complaint of right leg pain. Bone scan showed diffuse uptake of bone-seeking radiotracer on both lung fields predominantly in basal regions. The bronchoalveolar lavage test confirmed the diagnosis of PAM.

Keywords: Bone scan, pulmonary alveolar microlithiasis, tracer uptake

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a disease of unknown etiology marked by disseminated microliths (deposition of spherical calcium phosphate) throughout the lungs.[1,2] This disorder is an autosomal recessive disease and has currently been considered to be caused by mutation of a gene, which encodes a sodium phosphonate co-transporter.[1,2] The patient may complain progressive dyspnea; however, the diagnosis is often an incidental finding on chest X-ray radiography (CXR) or computed tomography (CT).[3] CXR and CT findings may be pathognomonic.[4] There is a mismatch between clinical and image findings in many cases. The symptoms and physical examination may be unremarkable while CXR and CT images may reveal diffuse involvement of lungs with dense reticulonodular opacities and calcified micronodules, bilaterally.[2]

After the disease is diagnosed, other family members of the patient should be screened by CXR.

The initial detection of the disease with other imaging modalities is rarely reported. ^{99m}Tc-diphosphonate scanning usually reveals diffuse intense uptake throughout both lungs.[3]

As to our knowledge <800 cases have been reported worldwide in the medical literature till date. About one-third of cases has been diagnosed in Asia. As well, one-third of cases are detected in Europe. Most of the cases have been reported from Turkey, Japan, and Italy. PAM occurs in both genders with a slightly higher incidence among males except in Italy where female predominance has been reported. Although, PAM may be detected in all age groups, it is most frequently diagnosed from birth to 40 years of age[5] with the maximum incidence in the third and fourth decades of the life.[2]

CASE REPORT

A 39-year-old man complaining a chronic progressive right leg pain was referred to our outpatient clinic on 28 July 2013 to perform a whole body bone scan. He denied any history of acute trauma.

At the first visit, physical examination was unremarkable. Both lower extremities were apparently symmetrical without any inflammatory sign or tenderness. There was no apparent neurological impairment or musculoskeletal abnormality in his legs. Range of motion for the knee joints and as well for ankle joints were in normal limits both in active and passive movements.

A whole body bone scan was performed 3 h after injection of 20 mCi ^{99m}Tc-methylene diphosphonate using a single-photon

Access this article online

Quick Response Code:

Website: www.ijnm.in

DOI: 10.4103/0972-3919.158548

Address for correspondence:
Dr. Bahar Moasses Ghafary, Research Center for Nuclear Medicine, Shariati Hospital, North Kargar Ave., 1411713135, Tehran, Iran.
E-mail: baharmoassesghafari@gmail.com

emission computed tomography/CT gamma camera equipped with low energy high-resolution collimator.

Whole body bone scan revealed a disseminated parenchymal uptake of the bone-seeking agent in bilateral lung fields [Figure 1a and b]. There was also increased radiotracer uptake in the right proximal tibiofibular joint, which was more likely due to an inflammatory and/or traumatic process [Figure 1c].

Based on the pulmonary abnormal findings on the bone scan images, a detailed history was taken more focused on the history of any pulmonary disease. He did not complain any severe pulmonary symptom except for somewhat intolerance to severe exercise. In addition, he mentioned that he had an abnormal CXR when he was 14 years old. His past medical history included an indolent subclinical pulmonary disease, which was not followed due to the stable X-ray findings and unremarkable clinical complaint.

A plain radiography, as well as CT, was obtained from his chest to characterize the parenchymal lung disease. Chest radiograph demonstrated a characteristic diffuse sand-like micronodulations (sand-storm appearance) throughout both lung fields [Figure 2]. CT of the chest also showed an extensive infiltrative dense calcified micronodulations in the parenchyma of both lungs [Figure 3]. The routine serum calcium and phosphorus laboratory profile was normal. The diagnosis of PAM was confirmed by fiber-optic bronchoscopy and bronchoalveolar lavage (BAL). Currently, the patient's brother is also suffering from PAM.

DISCUSSION

Pulmonary alveolar microlithiasis is characterized by intra-alveolar accumulation of spherical calcified nodules which are known as microliths. This disorder may involve people from any age ranging from early child to elderly.[6]

The proposed mechanisms include a hereditary error of metabolism, immune reaction to various irritants or infections, unusual response to pulmonary insults and acquired abnormality in the metabolism of calcium and phosphorous.[7]

The hallmark of PAM is incongruent clinical-radiological findings compatible with paucity of symptoms in relation to image findings.[6] The symptoms may be unremarkable even in the case of the extensive radiographic involvement. Some patients may present chronic cough and progressive dyspnea. Normal or mild restrictive pattern of pulmonary function tests may be present in asymptomatic cases 7.

Rarely, the cases are detected on bone scan images. Several mechanisms have been described for this finding; more accepted one is probably the same mechanism as described for bone uptake, that is, chemical absorptions on hydroxiapatite crystals. According to the X-ray micro-analysis, the microliths are formed by calcium, phosphate and a small amount of magnesium. The ratio of calcium to phosphorus indicates that the microliths may consist of hydroxiapatite-like materials, similar to that being detected in bone structure.[8]

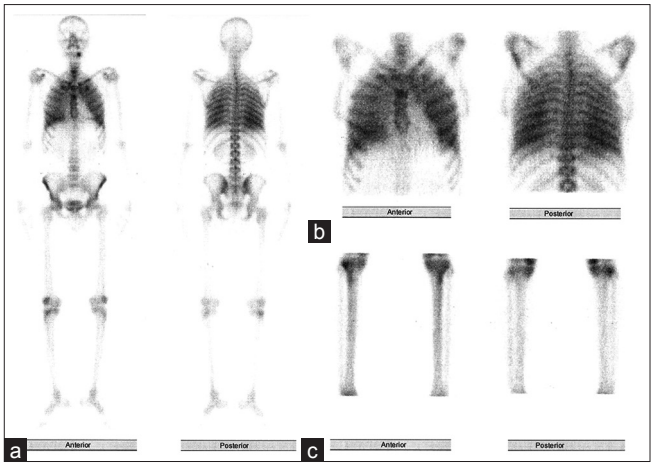

Figure 1: (a) Whole body bone scan. (b) Chest spot representing radiotracer uptake throughout both lung fields. (c) Legs spot reveals increased activity in the right tibiofibular joint

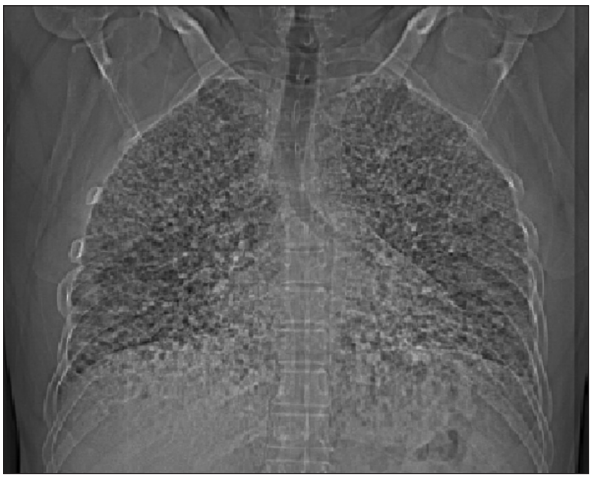

Figure 2: Chest X-ray demonstrated a characteristic diffuse sand-like micronodulations throughout both lung fields

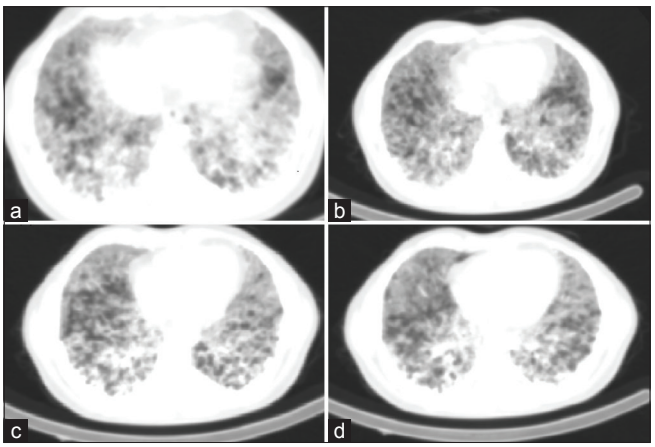

Figure 3: (a-d) Computed tomography of the chest showed an extensive infiltrative dense calcified micronodulations in the parenchyma of both lungs

Because the pattern of radiographic findings is typical and nearly pathognomonic,[4] other imaging and nonimaging modalities such as CT, BAL and transbronchial biopsy might be avoided in the presence of the typical radiological appearance.[3,9,10] The characteristic CXR findings in adults are ground-glass or fine sand-like or nodular pattern opacities that are more prominent in the middle and lower zones of the lungs. The calcification may be so dense that may wipe out the heart borders and the diaphragm. The most common CXR pattern in children is diffuse ground-glass opacities. Other less common findings are including bullae in the lung apical segments and/or plural calcifications.[7] The black plural line as a stripe of increased translucency between the lung parenchyma and the ribs are also described. These lines are often an early radiological finding in children and younger patients with PAM.[3,6,7]

The use of high-resolution CT (HRCT) allows the identification of minimal findings in the lung parenchyma.[9] HRCT may show thickening of the lobular septae in addition to the centrilobular distribution of microliths.[11]

In our patient, PAM was detected on the basis of diffuse pulmonary uptake of bone-seeking agent on bone scan images and the diagnosis was confirmed upon pathognomonic findings on CXR, CT and BAL.

Bone scintigraphy may also be useful in the detection of early small pulmonary calcified nodules, which have been associated with pulmonary dysfunction and cannot be identified by CXR due to the size smaller than the resolution of conventional radiograph images.[6] The bone scan in our case shows bilateral extensive parenchymal uptake of the bone-seeking agent. Unfortunately, no definitive therapy existed for PAM. Lung transplantation may be useful in special cases with severe impairment of the pulmonary function.[12]

CONCLUSION

As a result, PAM is a rare idiopathic lung disease representing calcified micronodules in bilateral lung parenchyma. Radiographic findings are pathognomonic in most cases of PAM. Bone scintigraphy often shows diffuse bilateral uptake of radiotracer throughout the lung fields, especially in early phases of the disease when no typical CXR findings are detected.

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How to cite this article: Fallahi B, Ghafary BM, Fard-Esfahani A, Eftekhari M. Diffuse pulmonary uptake of bone-seeking radiotracer in bone scintigraphy of a rare case of pulmonary alveolar microlithiasis. Indian J Nucl Med 2015;30:277-9.

Source of Support: Nil. **Conflict of Interest:** None declared.