Case Report

Role of ^{99m}Tc-MDP bone scan in the diagnosis of Erdheim–Chester disease

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ABSTRACT

Erdheim–Chester disease (ECD) is a rare systemic non-Langerhans cell histiocytosis. It is a progressive disease of unknown etiology. The ^{99m}technetium-methylene diphosphonate (^{99m}Tc-MDP) bone scan is useful in finding the sites of involvement in the skeleton and is helpful in excluding other causes of bony pain. Also a scintigraphic pattern consistent with ECD should alert the physician to evaluate the patient for visceral sites of involvement using fludeoxyglucose positron emission tomography/computed tomography (FDG PET/CT), as this is known to be fatal at times.

Keywords: Erdheim chester disease, fludeoxyglucose positron emission tomography/computed tomography, ^{99m}Tc MDP bone scan

INTRODUCTION

Erdheim–Chester (ECD) disease is a rare systemic non-Langerhans cell histiocytosis first described by William Chester in 1930.[1] Jaffe in 1972 first coined the eponym Erdheim–Chester disease.[2] The disease affects both males and females, though with a slight predilection for men, and has a peak incidence between the fifth and sixth decade of life, although manifestation can be at any age.[3] ECD is a progressive disease of unknown origin that has a 5-year survival rate of 41%.[4] It is not known definitely if ECD belongs to the histiocytosis family or is a distinct entity;[5] however, some authors have categorized ECD as a lipid storage disease[6] or as a primary disorder of the monocytes and macrophages.[7] However, on immunohistochemistry the infiltrating cells stain positive for CD68 and negative for CD1a and S-100.[8]

CASE REPORT

A 65-year-old male patient presented with complaints of bone pain at multiple sites and bilateral exophthalmos. The patient underwent serum calcium and alkaline phosphatase level evaluation, which was 8.7 mg/dl and 80 IU/l (normal value 20-140 IU/l), respectively. The patient underwent a ^{99m}technetium-methylene diphosphonate (^{99m}Tc-MDP) three phase skeletal scintigraphy, which revealed increased activity in bilateral orbital region (right > left) in flow and blood pool phases, whereas diffusely increased tracer uptake in the skeleton was noted with high bone to soft tissue contrast and faintly visualized kidneys. Along with this, intense tracer uptake was noted in bilateral orbits, nasal bones, and medial part of bilateral maxillae [Figure 1]. Axial section through the orbit in a non-contrast CT showed bilateral retrobulbar soft tissue density masses [Figure 2]. A non-contrast CT done elsewhere, through bilateral upper femora and pelvis showed multiple osteosclerotic foci. Fine needle aspiration cytology performed from the skeletal lesion revealed xanthogranulomatous infiltration by foamy histiocytes surrounded by fibrosis. A provisional diagnosis of histiocytosis was made. Further immunohistochemistry to characterize the type returned positive for CD68 and negative for CD1a and S-100. Based on the clinical, scintigraphic, and histopathological findings, a diagnosis of ECD disease was made. Because ECD often involves the kidneys, the patient was evaluated for the same, which revealed urea and creatinine levels of 36 and 0.8 mg/dl, respectively.

DISCUSSION

Patients with ECD most commonly present with bone pain mainly in the lower limbs.[9] The classic imaging finding is bilateral and symmetric mixed cortical sclerotic and lytic lesions in the metaphyseal and diaphyseal regions of the long tubular bones

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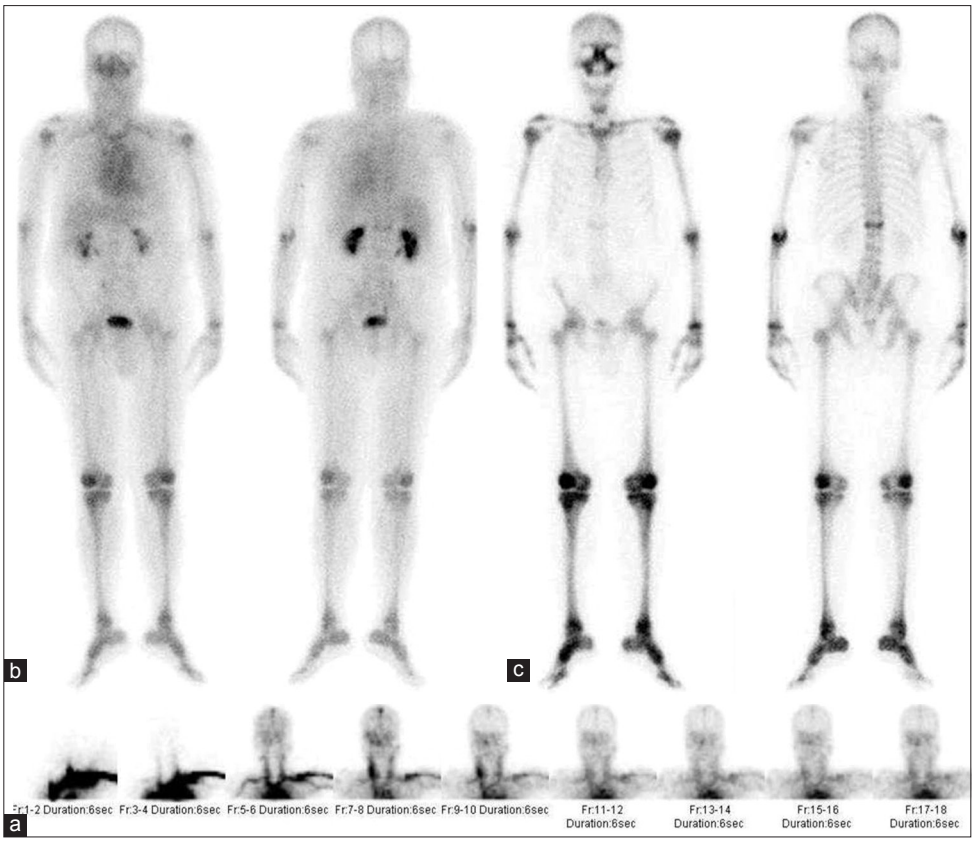

Figure 1: (a) The ^{99m}Tc-MDP bone scan-flow phase shows increased blood flow to bilateral orbital regions (right > left). ^{99m}Tc-MDP = ^{99m}technetium-methylene diphosphonate (b) The ^{99m}Tc-MDP bone scan-soft tissue (pool) phase shows increased activity in bilateral orbital/infraorbital regions (right > left). Also seen is increased activity in L1 vertebra region in a linear horizontal pattern. Mild increase in activity is noted diffusely in the axial and appendicular skeleton. ^{99m}Tc-MDP = ^{99m}technetium-methylene diphosphonate (c) The ^{99m}Tc-MDP bone scan-delayed phase whole body anterior and posterior images show diffusely increased tracer uptake in the skeleton with high bone to soft tissue contrast and faintly visualized kidneys. Also noted is intense tracer uptake in bilateral orbits (especially inferior and medial walls), nasal bones, and medial part of bilateral maxillae. Appendicular skeleton reveals overall increased uptake. The L1 vertebra shows a linear horizontal increased uptake due to compression/osteoporotic collapse. ^{99m}Tc-MDP = ^{99m}technetium-methylene diphosphonate

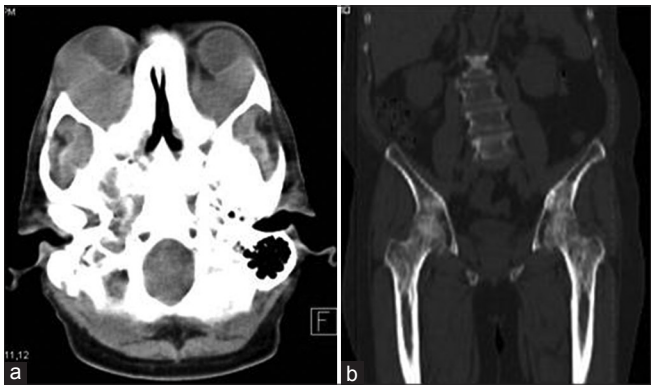

Figure 2: (a) Axial section through the orbit in a non-contrast CT showing bilateral retrobulbar soft tissue density masses. (b) Coronal section of a non-contrast CT through bilateral upper femora and pelvis showing multiple osteosclerotic foci

with sparing of the epiphyses and axial skeleton.[3,4,8,10] Besides this characteristic skeletal involvement, patients with ECD may show a 'hot' kidney on radionuclide bone imaging, which may imply renal parenchymal involvement by disease.[11] Approximately 50% of patients present with extraskeletal manifestations; the most commonly involved sites being the heart, the lungs, the kidneys, the retroperitoneal space, the central nervous system, and the skin.[4]

The most frequent central nervous system manifestation is central diabetes insipidus followed by cerebellar symptoms, usually ataxia of gait.[12]

Clinical course depends on disease extension and distribution: Central nervous system and cardiac involvement are associated with poor prognosis.[13] As treatment of ECD is difficult, it is important to know the extent of the disease.[14]

Our case shows the importance of a bone scan in the diagnosis of this condition, as its diagnosis is of utmost importance. This can guide the clinician to look for other sites of involvement using fludeoxyglucose positron emission tomography/computed tomography (FDG PET/CT), as pulmonary and myocardial involvement can be fatal at times.

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