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99mTc-MDP bone scan in Erdheim-Chester disease: case description

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CASE DESCRIPTION

A 54-year-old male patient complaining of bone pains, dyspnea, and bilateral exophthalmos for the last 2 years was referred to the nuclear medicine department for a bone scan. His contrast-enhanced magnetic resonance imaging brain and orbit showed bilateral symmetrical T2WI hypo intense retro-bulbar lesions with homogenous post-contrast enhancement. Microscopic appearance and immunohistochemistry findings of orbital biopsy favored xanthogranuloma. ^{99m}Tc MDP bone scan showed an abnormally increased radiotracer uptake in the region of the ethmoid and bilateral maxillary sinuses along with an abnormally increased uptake in a diffuse manner involving humerus, radius ulna, tibia, and ends of femur bilaterally (Fig. 1). Correlative single photon emission computed tomography/computed tomography (CT) showed bilateral retro-bulbar soft tissue density lesions causing proptosis (Fig. 2A), mucosal thickening in maxillary sinuses (Fig. 2B), and left-sided pleural effusion (Fig. 2C). Clinical, histopathological, and typical scintigraphic patterns favored the Erdheim-Chester disease (ECD).

ECD is a rare systemic non-Langerhans cell histiocytosis characterized by the infiltration of foamy lipid-laden histiocytes in tissues [1]. It has slight male predominance and can manifest at any age with a peak incidence between the fifth and sixth decade of life [2]. Almost all patients diagnosed with ECD have some degree of bone involvement (95%); however, sites of the extraosseous disease include the sinuses, large vessels, retroperitoneum, heart, lungs, central nervous system, skin, pituitary gland, and orbits [3].

Bone pain is the first revealing symptom in most of the patients. The osseous involvement in ECD is characterized by symmetrical sclerosis of the metaphysis and diaphysis of the long tubular bone bilaterally with sparing of the epiphyses and axial skeleton [2]. However, due to the systemic involvement of ECD, its imaging findings can be highly variable.

The ^{99m}Tc-MDP bone scan is useful in determining the extent of this rare disease in the skeleton. The familiarity of physicians with the typical scintigraphic pattern of ECD is a prerequisite to warrant further imaging such as fludeoxyglucose positron emission tomography CT to look for visceral involvement.

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Declarations

Conflict of interests

The authors declare that they have no conflict of interest regarding the publication.

Funding

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Consent to participate

Informed consent was obtained from the patients.

Ethics approval

Ethics approval is not required at our institution for publishing this type of manuscript in a medical journal.

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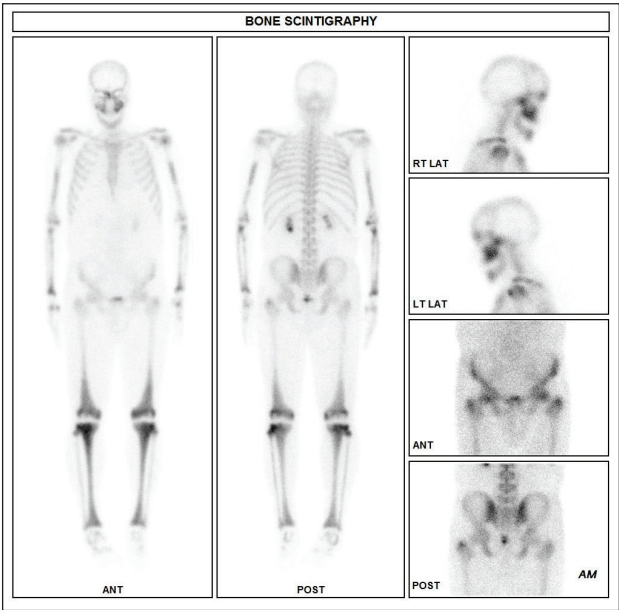


Figure 1. 99mTc-MDP bone scan.

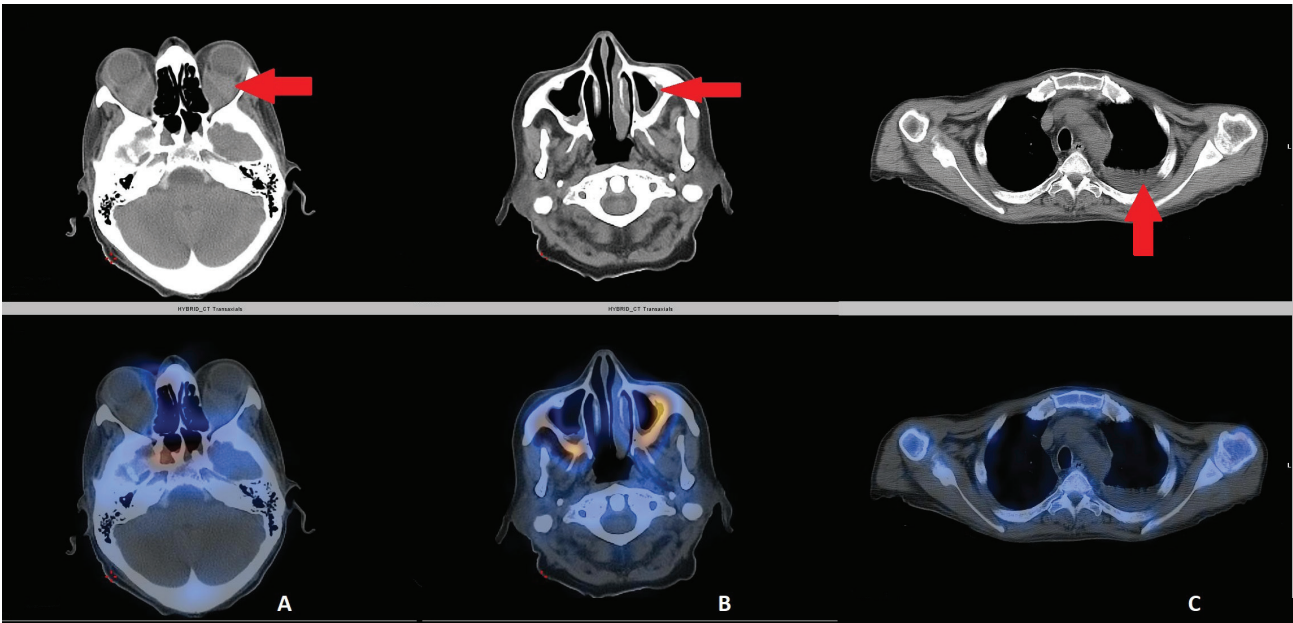


Figure 2. SPECT/CT showed bilateral retro-bulbar soft tissue density lesions causing proptosis (A), mucosal thickening in maxillary sinuses (B), and left-sided pleural effusion (C)(marked with red arrows).