Correspondence

Erdheim-Chester’s disease as a differential diagnosis of “hot” kidneys on bone scintigraphy

To the Editor: We read with interest a case of Erdheim-Chester disease (ECD) published in HJNM 2008; 10: 164-167 [1] and we would like to present another case which differs from the above as having an unusual bone involvement and “hot” kidneys on bone scintigraphy. The patient was a 46 years old man admitted for evaluation of the pain that he had in his lower limbs for the last 3 years. He also had weakness, weight loss and diabetes insipidus. Physical examination revealed pitting edema of the lower limbs and some cutaneous xanthelasmata. Serum creatinine was normal. Sonography of the kidneys demonstrated increased renal size (145×67×28mm for left kidney and 140×66×24mm for right kidney) and some corticomedullary loss of image differentiation without evidence of obstructive calyceal dilatation. X-rays of both proximal and distal femora showed symmetric metaphyseal and diaphyseal involvement of lesions including mixed osteosclerosis and lytic areas (Fig. 1). Bone scintigraphy with technetium-99m-methylene diphosphonate (99mTc-MDP) revealed multiple bone involvement (Fig. 2 and 3). Sites of symmetrical increased radionuclide uptake included humeri, scapulae, radii, femori, tibiae, tarsal and metatarsal bones. Right iliac bone also showed focal hyperactivity. The skull and the vertebral column were intact. Furthermore, both kidneys demonstrated markedly increased radionuclide uptake (Fig. 2 and 3). The patient had not taken any nephrotoxic drugs before or during our examination.

Bone biopsy from right femoral lateral epicondyle showed fibro-collagenous and fatty tissue infiltrated by clusters of foamy histocytes with central vesicular nuclear and abundant vacuolated cytoplasm. Some Touton-shape giant cells were noted. There was also small aggregation of histiocytic like cells with eosinophilic cytoplasm and ovaloid nuclei. Renal biopsy demonstrated similar parenchymal infiltration. These pathologic findings supported the diagnosis of ECD [1].

Erdheim-Chester disease is a disseminated xanthogranulomatous disease of different organs and bones that are infiltrated by foamy histocytes [1]. It remains unclear whether ECD belongs to the histiocytosis family or is a distinct entity [2], however some authors categorized ECD as a lipid storage disease [3] or as a primary disorder of the monocytes and macrophages [4]. One hundred sixty cases have been reported up to 2004 [5]. Bone pain is the most frequent reported symptom, mostly located in the lower limbs [6]. Exophthalmos, diabetes insipidus, general symptoms (fever and weight loss) and xanthomas are also clinical findings of the disease [6]. Extraskeletal involvement of central nervous system, heart, pericardium, lung, pleura, retroperitoneum, liver, spleen, kidney, breast, gingiva, conjunctiva and orbits have been reported [6]. Symmetrical long bone osteosclerosis is the radiological sign specific for ECD. Langerhans cell histiocytosis (LCH) can have some overlapping clinical and histological findings, however ECD may differ from LCH by immunohistologic and microscopic characteristics. In contrast, skeletal lesions of LCH are osteolytic and very rarely located in the long bones [7]. Because of histological findings, some authors call ECD as polyostotic sclerosing histiocytosis [8]. This widespread sclerosis of the appendicular skeleton is rarely associated with lytic lesions and usually spares the
epiphysis and axial flat bones. Differential diagnosis includes mastocytosis, fluoride intoxication, myeloid metaplasia, lymphoma, metastatic disease, toxic osteoarthropathy and adult progressive diaphyseal dysplasia (Englemann disease) [9]. Although epiphyseal involvement is rarely reported [9], our case demonstrated increased radionuclide uptake in epiphyses as well as in metaphyses and diaphyses. Bone scanning showed skeletal involvement in the bones of the lower extremities.

Renal and perirenal involvement is found in about 29% of the cases [10]. Kidneys are frequently abnormal on imaging or necropsy, but clinical manifestations like abdominal pain, enlarged palpable kidneys and obstructive renal impairment are uncommon [11]. Our case demonstrated bilaterally marked renal radionuclide uptake resulting from bilateral renal parenchymal involvement. In addition, there was an unusual asymmetry in size of both kidneys that could be due to different involvement of each kidney. To the best of our knowledge, there are few reported cases of ECD that had significantly high renal parenchymal uptake of the bone imaging agent, although nephromegaly and “hairy kidney” appearance on an abdominal CT have been reported [12]. Thus ECD should be considered in the spectrum of differential diagnosis of “hot” kidneys on radionuclide bone scanning. In addition, “hot” kidneys may imply renal parenchymal involvement during disease progression.

In conclusion, besides other typical bone scan findings, Erdheim-Chester disease should be considered in the spectrum of differential diagnosis of “hot” kidneys on bone scintigraphy.

Bibliography


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