Intraosseous Hemangiomatosis: Technetium-99m(V)Dimercaptosuccinic Acid and Technetium-99m-Hydroxymethylene Diphosphonate Imaging

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We report a case of histologically proven intraosseous hemangiomatosis in which marked accumulation of pentavalent technetium-99m-dimercaptosuccinic acid (^{99m}Tc(V)DMSA) and technetium-99m-hydroxymethylene diphosphonate (^{99m}Tc-HMDP) was observed in the osteolytic hemangiomatous lesions.

Key Words: intraosseous hemangiomatosis; bone scintigraphy; technetium-99m-hydroxymethylene diphosphonate; pentavalent technetium-99m-dimercaptosuccinic acid

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Intraosseous hemangiomatosis, or Gorham's disease, is a rare skeletal disorder of unknown etiology representing a syndrome of massive osteolysis or so-called "disappearing bone disease" (*1–3*). No bones are spared by this disease. Radiographically, the disease is characterized with multiple osteolytic lesions (*4*) which progress and eventually lead to deformity and shortening of extremities (*1*). Pathological fracture is an uncommon complication and often requires surgical treatment.

Radiographic and bone scintigraphic findings in patients with massive osteolysis have been described (*5,6*). However, scintigraphic findings using a tumor-seeking radiopharmaceutical have not yet been reported. Here we report a case of intraosseous hemangiomatosis where marked accumulation of ^{99m}Tc(V)DMSA and ^{99m}Tc-HMDP in bone lesions was observed.

CASE REPORT

A 35-yr-old male with chronic pain in the left upper arm was previously diagnosed as having an osteolytic lesion at the distal end of left humerus in 1961. Similar osteolytic lesions developed thereafter in bilateral upper limbs, ribs, bilateral distal femurs, proximal tibias and the right scapula. Open biopsy was performed four times at different sites and revealed cavernous hemangioma of the bone. In 1985, a pathological fracture occurred in the deformed and shortened right humerus. Surgery with a specially designed total elbow replacement was performed. In 1988, the

FIGURE 1. Plain radiogram of the right knee shows widespread osteolytic lesions with slight expansion of the cortex at distal half of the femur and proximal half of the tibia.

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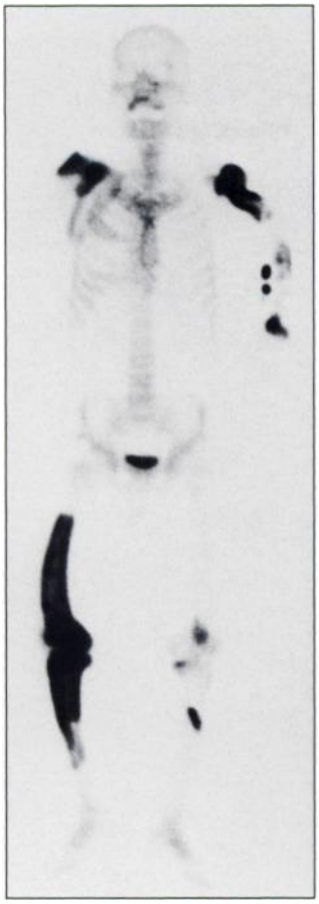

FIGURE 2. Technetium-99m-HMDP bone scintigraph shows markedly high accumulation at right leg, right scapula, right acromion, left shoulder, left wrist, left knee, left lower leg and left elbow and faint uptake in right third rib. The marked uptake at left elbow was concordant with the site of loosened intramedullary stem of the elbow prosthesis in the radius and ulna for the surgical treatment of pathologic fracture.

FIGURE 3. Tumor-seeking scintigraphy using ^{99m}Tc(V)-DMSA revealed markedly abnormal uptake in right lower leg, right scapula, right acromion, left shoulder and left wrist and faint uptake in left elbow and knee. No evident accumulation was found at the left elbow in which evident uptake was found on the bone scintigram.

right upper limb was amputated after the patient developed osteosarcoma in the right radius.

Serum biochemical parameters including serum calcium and phosphate levels were normal, except for the serum alkaline phosphatase (types 2 and 3) which was persistently elevated.

Skeletal x-ray examination showed multiple osteolytic lesions in bilateral limb bones, right scapula and ribs (Fig. 1). Bone scintigraphy using ^{99m}Tc-HMDP showed marked accumulation of the tracer in the osteolytic lesions. In addition, intense uptake of the tracer was observed in the regions adjacent to the intramedullary stem of the elbow prosthesis in the left radius and ulna (Fig. 2). Scintigraphy using ^{99m}Tc(V)DMSA, a tumor-seeking agent (*7*), revealed marked tracer uptake exclusively in the osteolytic lesions, and not in the regions surrounding surgical prosthesis (Fig. 3).

DISCUSSION

This report compares ^{99m}Tc(V)DMSA imaging with ^{99m}Tc-HMDP bone scanning in a patient with intraosseous hemangiomatosis. We have previously shown that ^{99m}Tc(V)DMSA, a ^{99m}Tc-labeled tumor-seeking agent, is useful to detect a variety of progressive and hypervascular softtissue tumors, including low-grade sarcomas (*7*), aggressive fibromatosis (*8*), tenosynovial giant cell tumors (*9*) and amyloid tumors (*10*), in which ^{67}Ga scanning is negative.

The bone scan of this patient is similar to that in a case of Paget's disease and acro-osteolysis involving long bones and that in a case of fibrous dysplasia involving the ribs and the scapula. The distribution of the lesions and their radiolucent and expansile features on plain radiograms can easily exclude these diseases (*11*).

Markedly enhanced ^{99m}Tc(V)DMSA uptake was found only in the osteolytic bone lesions, and not the regions surrounding the loosened surgical prosthesis in the left radius and ulna (Fig. 3). This finding contrasts with the results of ^{99m}Tc-HMDP imaging in which increased tracer accumulation was observed in the regions surrounding the loosened surgical prosthesis in the left radius and ulna in addition to the primary osteolytic bone lesions. Although the mechanism of ^{99m}Tc(V)DMSA accumulation in the tumor tissue remains unknown (*12*), the accumulation pattern of ^{99m}Tc(V)DMSA appears tumor-specific and may provide a useful clinical tool for direct evaluation of the skeletal hemangiomatous lesions on initial and follow-up examinations.

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