
Nuclear Medicine Imaging of Systemic Mastocytosis

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A 66-yr-old male with systemic mastocytosis is presented along with classic imaging findings of dense bone on radiographs, intense uptake on bone scan and marrow expansion on bone marrow scan. Dual photon absorptiometry (DPA) measurements of the skeleton revealed a very high bone mineral measurement of 1.678 grams per square centimeter (lumbar vertebrae 2 through 4). It is suggested that DPA may be useful to follow the development of mast cell disease in the bones.

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Systemic mastocytosis is an unusual disorder that is characterized by a proliferation of mast cells that infiltrate the bone marrow, spleen, liver, skin, and lymph nodes. (1) The disease has infiltrative and pharmacologic manifestations. The characteristic skin rash, urticaria pigmentosa, is a result of accumulations of mast cells in the skin. Lymph nodes, spleen, and liver may be enlarged by mast cell infiltration. The bone marrow may be occupied by increased numbers of mast cells. Release of histamine, serotonin, and mediators of immediate hypersensitivity lead to fever and flushing. Diarrhea, weight loss, and peptic ulcer disease are also clinical manifestations. Focal or diffuse sclerotic, dense bones or osteopenia may be seen by radiographic examination. Liver-spleen scanning can demonstrate organomegaly. Bone scanning has been reported to show patchy or diffusely increased tracer uptake. Bone marrow scanning may document an expanded distribution of colloid (2). Bone mass measured by dual photon absorptiometry (DPA) has to our knowledge not been reported in systemic mastocytosis.

CASE REPORT

A 66-year-old white man had urticaria pigmentosa documented by biopsy in 1983 (Fig. 1). His complaint of chronic diarrhea and recent peptic disease (documented by endoscopy)

were notable clinical features at the time of his imaging evaluation. The patient complained of episodic flushing and pruritus of recent onset. Radiographs showed diffuse, dense bones with thickened trabeculations (Fig. 2). There was intense uptake of technetium-99m methylene diphosphonate (^{99m}Tc) MDP in the entire skeleton with faint renal excretion. An unusually high bone to soft-tissue ratio was qualitatively observed (Fig. 3). A bone marrow scan established an extended reticuloendothelial system and moderate splenomegaly (Fig. 4). A DPA (Lunar -DP4) examination of the first through fourth lumbar vertebra showed a marked increase in bone mass (Table 1). A diagnosis of systemic mastocytosis was made.

DISCUSSION

Approximately 90% of the cases of this mast cell disease are limited to the cutaneous form with urticaria pigmentosa. The skin disease coupled with bone changes is the second most common presentation. As many as 10% of systemic mastocytosis cases have clinical manifestations referable to involvement of bone, liver, spleen, or lymph nodes. In rare cases some form of leukemia will ultimately develop (3-5).

Urticaria pigmentosa occurs most often in young children and tends to regress in adolescents. Approximately 25% of cases of recognized mast cell disease begin in adulthood. The bone lesions of systemic mastocytosis are particularly common when the onset of the disease is after the age of 50 yr as in the case discussed (3). The adult cases tend to progress to a more severe disease. The most common pattern is that of diffuse demineralization. Osteosclerosis is also com-

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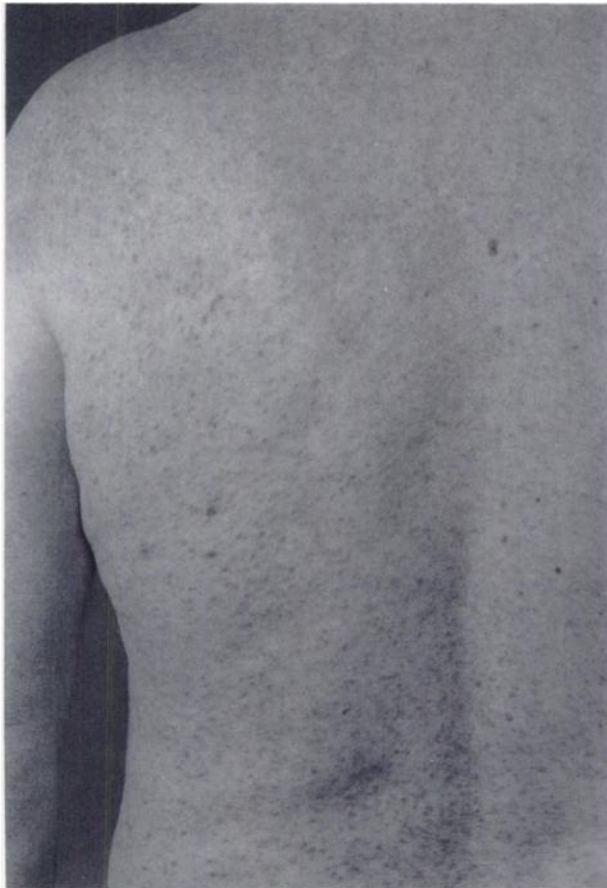


FIGURE 1
The characteristic *urticaria pigmentosa* skin rash seen on the back. It was also present on the arms and chest.

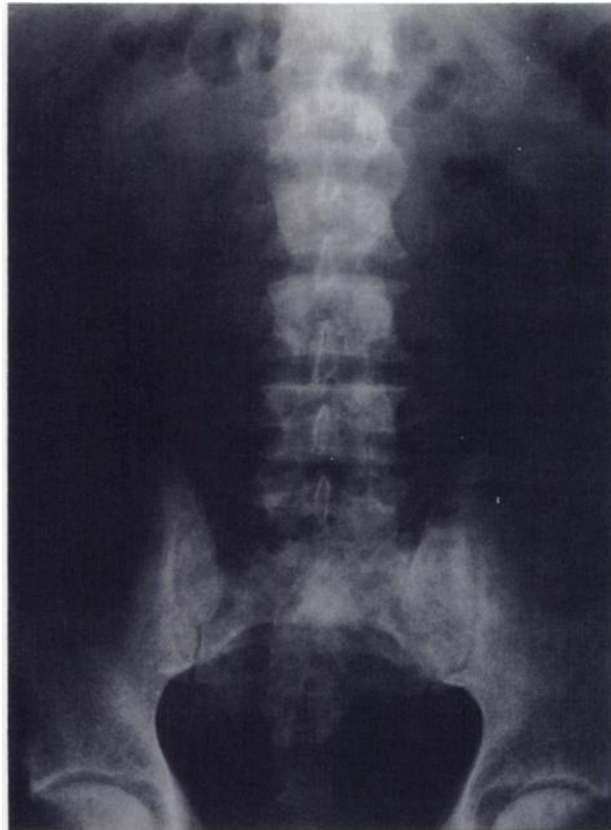


FIGURE 2
A radiograph of the lumbar vertebrae shows dense bone which on close inspection is comprised of coarse trabeculations.

mon. Production of heparin by the mast cells is believed to be partly responsible for osteopenia. Sclerosis is not seen with heparin induced bone disease. Other mast cell products likely contribute to the sclerotic bone changes seen in mastocytosis. The mechanism is incompletely understood. Pathologic fractures occur in a minority of patients (5).

Radionuclide scans (marrow and bone) are more sensitive than radiographs for the detection of skeletal involvement in systemic mastocytosis. Bone scans range from normal to unifocal, multifocal and diffusely abnormal in patients with this illness. The highest mast cell burdens correspond to the more positive images (6). Diffusely increased skeletal uptake in the pattern of a "superscan" is known to develop in conjunction with hematologic diseases that have expanded marrow space (7). Reticuloendothelial expansion associated with marrow expansion in myeloproliferative disorders can be readily imaged with colloid scans (8). None of these imaging tests are specific for the disease but may help exclude systemic involvement.

The DPA measurements are likewise not specific but offer an accurate way to track changes in bone density

(9). The vast majority of scientific communications addressing DPA are in reference to bone mineral loss but some endocrinologic diseases such as hypoparathyroidism and acromegaly have been found to have higher than normal bone mass (10). DPA is not needed to diagnose systemic mastocytosis but could easily be used to track the bone involvement and to study the natural history of this disease. Perhaps radiographically undetected bone mass abnormalities could be documented when the disease is at an earlier stage.

TABLE 1
DPA Data

Region	Grams/CM ²	% Age-matched ^a	Z-score
L-1	1.397	138.6	3.4
L-2	1.716	139.2	3.7
L-3	1.750	142.0	4.0
L-4	1.602	130.0	2.8
L2-4	1.678	136.1	3.4

^a 66-yr-old white male, height 180 cm, weight 95.9 kg.

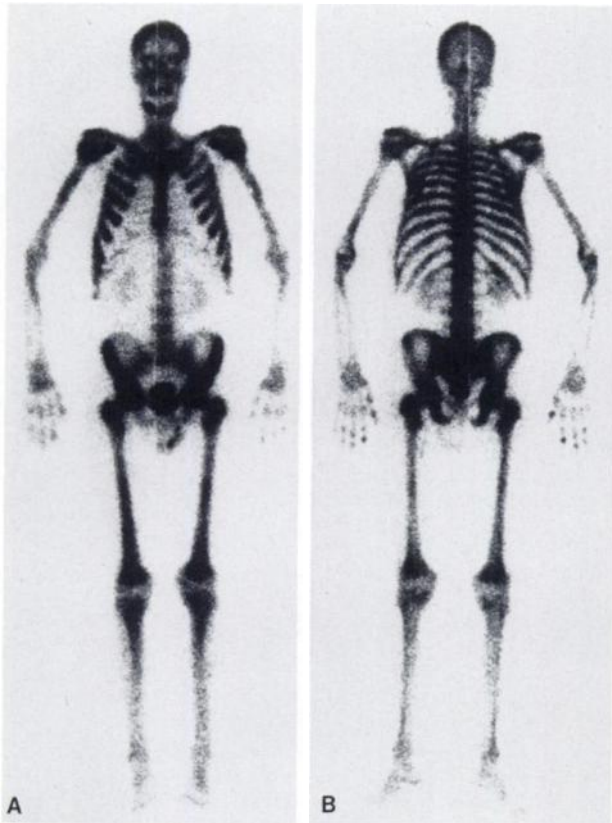


FIGURE 3
An anterior (A) and posterior (B) whole-body scan done 3 hr after injection of 20 mCi [^{99m}Tc]MDP is nearly a "super-scan" with intense bone labeling and faint renal activity.



FIGURE 4
An anterior bone marrow scan done with 10 mCi of [^{99m}Tc] albumin colloid documents the expansion of erythropoietic marrow into the extremities. Note the enlarged liver and spleen.

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