# Bone Scan in Mastocytosis: Case Report

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***A 45-year-old man with well-documented systemic mastocytosis showed generalized symmetric increased activity on bone imaging. These scan findings are grossly indistinguishable from those of patients with renal osteodystrophy or secondary hyperparathyroidism. The images of the hands, however, failed to show the changes observed in secondary hyperparathyroidism. The mechanism for this intense activity is thought to be due to aberrant new-bone formation.***

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Mastocytosis, or urticaria pigmentosa, is an uncommon disease. Although it was once regarded as a purely dermatologic entity, systemic involvement was later recognized (*1,2*). In 1952, Sagher et al. (*2*) first described the osseous changes on radiographs in a case of mastocytosis. Subsequently, additional examples with bony changes were recorded radiographically (*3–5*).

This communication describes the abnormal bone-scan findings in a man with a long-standing history of mastocytosis. The images, at 5:1 minification, were obtained with an Ohio-Nuclear Model-84 scanner, 3 hr after the intravenous injection of $^{99m}$Tc-disodium etidronate. To our knowledge, the skeletal findings on scintillation imaging in this entity, although probably nonspecific, have not previously been described.

## CASE REPORT

A 45-year-old man was admitted with an established diagnosis of systemic mastocytosis. Before adolescence, he had suffered intermittent attacks of wheezing and urticaria, sometimes precipitated by exposure to dust, dandruff, or goose feathers or by the ingestion of spinach, carrots, or pork. At the age of 17, brown spots were noticed on his arms, trunk, face, and lower extremities. In 1954, a biopsy of the skin revealed the classic histologic features of mastocytosis. The characteristic Darier's sign of urticaria pigmentosa was first seen in the patient at that time. This dermatographia has persisted through the years. Between 1960 and 1975, the patient experienced numerous attacks involving flushes, weakness, palpitations, nausea and vomiting, severe hives, and loss of consciousness. The symptoms varied in intensity.

Physical examination revealed numerous macular and maculopapular brownish-red skin lesions of variable size and shape, heavily involving the face and trunk, somewhat less dense on the upper extremities and least prominent on the lower extremities. Darier's sign could be elicited in any area of integument spotted with pigment. The examination of other systems was not otherwise remarkable.

**Laboratory examinations.** A complete blood count was normal except for hypersegmentation of the polymorphonuclear neutrophils. Bone marrow biopsy revealed an increased count of mast cells, with most of them located around marrow spicules. Limited trabeculae appeared thickened. Except for some elevation of the serum alkaline phosphatase level, the remainder of the SMA-12 study was normal. A skull x-ray was normal. A skeletal survey showed increased bone density with cystic changes in the medullary cavity. Endosteal thickening and localized areas of bone sclerosis were noted, especially in the proximal left humerus and in several ribs. Skin biopsy was reported as mastocytosis.

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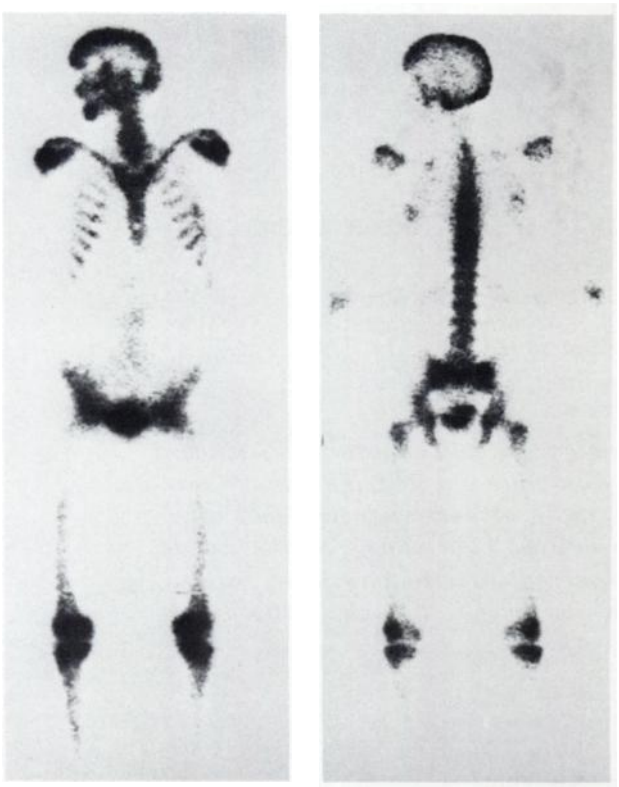

**FIG. 1.** Symmetric heavy concentration of activity in axial skeleton and skull, mandible, and appendicular bones in scan with $^{99m}$Tc-disodium etidronate. Note decreased uptake in kidneys (absent kidney sign).

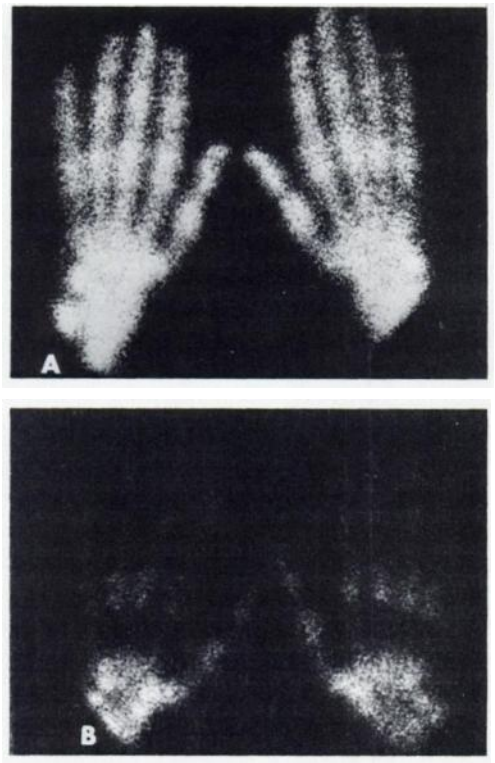


**FIG. 2.** (A) Intense activity in phalanges and interphalangeal joints in patient with secondary hyperparathyroidism. (B) Normal hand images in patient with mastocytosis.

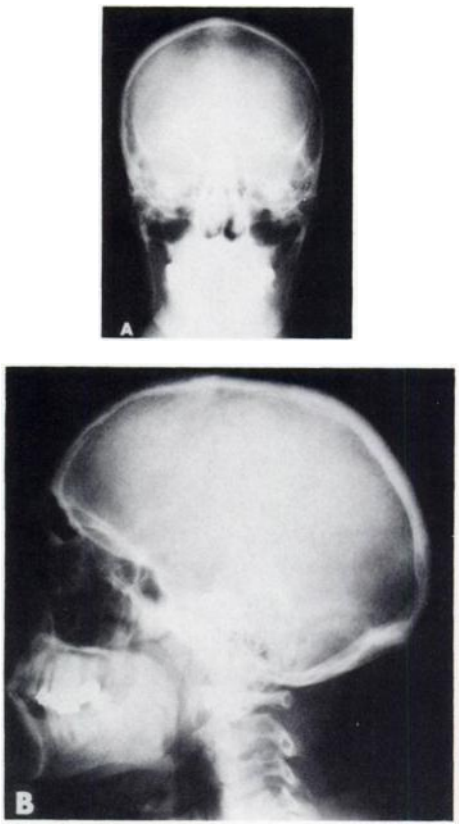


**FIG. 3.** Normal skull x-rays in patient with mastocytosis.

## DISCUSSION

The bone scans show pronounced activity, symmetrically distributed in both the axial and appendicular bones (Fig. 1). The large long bones showed the most activity at the ends, although the shafts of the femur and tibia also showed moderate uptake. The clavicles, individual ribs, and vertebrae are well resolved because of the enhanced uptake of tracer. The observed decrease in kidney uptake ("absent kidney sign") also suggests an unusually heavy bone concentration. This sign has also been recorded in instances of widespread bony metastases (*6*) and severe Paget's disease (*7*).

The generalized distribution of increased radioactivity does not appear to be distinguishable from the pattern observed in those patients undergoing chronic hemodialysis who manifest severe renal osteodystrophy or from the bony changes due to hyperparathyroidism (*8*). The mechanism for the generalized intense activity in our patient is probably related to the aberrant new-bone formation in mastocytosis, which usually results in thickened trabeculae of the spongiosa and thickened cortex of the long bones with narrowing of the marrow space and generalized osteosclerosis (*3*). The increased bone density and localized areas of sclerosis on the radio-

graphs do tend to suggest this. In our patient, however, the phalanges of the hands and periarticular areas do not show any abnormal uptake, in contrast to the well-outlined phalanges and interphalangeal joints observed in patients with proven secondary hyperparathyroidism (Figs. 2A and 2B). We are not certain whether this difference will prove sufficient to distinguish these entities.

Although the patient's history of mastocytosis dates back at least 28 years, when the skin manifestations first surfaced, the skeletal x-rays showed limited changes only. The extent of the bony involvement appears to be better suggested by the bone scans, especially in the calvarium, where no x-ray changes were evident (Figs. 3A and 3B). The cystic changes in some of the ribs and left humerus, however, were not seen on the scintigrams as such, possibly because of the small size of the lesions.

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