

CASE REPORTS

Bone and Gallium Scans in Mastocytosis: Correlation with Count Rates, Radiography, and Microscopy

Richard D. Ensslen, Frank I. Jackson, and Anne M. Reid

Cross Cancer Institute, Edmonton, Alberta, Canada, and Hinton General Hospital, Hinton, Alberta, Canada

Mastocytosis (urticaria pigmentosa) was proven in a patient suffering from severe back pain. A bone scan showed diffusely increased bone activity. Count rates were also abnormally elevated over several areas of the skeleton. Radiographs were consistent with mastocytosis in bone.

J Nucl Med 24: 586-588, 1983

Mastocytosis was first described in the dermatologic literature. Later, systemic involvement was recognized (1-4). Radiographic changes of the skeleton were first described by Sagher in 1952 (5). Subsequently, additional radiographic involvement of bone has been noted (1,6-9).

Increased activity on the bone scan was first described in 1976 by Sy in one patient (10), later by Sostre in another (11), and by Deramond in a third (12).

We present here a patient with radiographic, scintigraphic, and electron-microscope findings.

CASE REPORT

An 83-yr-old woman was admitted to hospital for investigation of severe back pain in the lower dorsal and upper lumbar regions. Radiographs were reported as "highly suggestive of metastatic bone disease" (Fig. 1). The patient's pain settled on rest and meperidine, and she was referred for assessment and a search for a primary.

The radiographs were reviewed (F.I.J.) and the differential diagnosis of metastatic disease to bone, or alternatively, mastocytosis was made independently of the history.

Whole-body planar tomographic bone scans were made with Tc-99m methylene diphosphonate (Tc-99m MDP), 189 μ Ci (7 MBq) per kg of body weight (Fig. 1). The instrument* yields 12 coronal slices of the whole body. Diffusely increased bony activity was demonstrated, this being particularly striking in view of the patient's age. The increased activity in the appendicular skeleton suggested a hematological disorder or a hypermetabolic state. The

differential diagnosis of a "superscan" was also entertained (13-18).

Our laboratory routinely obtains counts over a number of skeletal sites† whenever a bone scan is done on a patient with carcinoma of the prostate or breast, and at times in unusual bone scans such as in this case. Our upper limits of normal usually are 85-90 K counts/min when the scan is done 3¼ hr after injection, but the count rates are normally much lower in a patient of this age.

The quantitative part of the study confirmed the increased activity seen on the scan, and also showed that the process was locally variable. In particular, much less activity was seen in the sternum and cervical spine than in other measured skeletal areas (Table 1).

The increased skeletal activity and the nature of its distribution were considered compatible with the suspicion of mastocytosis.

The patient's laboratory screen showed normal CBC, urinalysis, and electrolytes. Only two abnormal biochemical results were found: an elevated alkaline phosphatase of 145 (N: 15-65 IU/l), which has been described in this disorder (10,19), and a raised uric acid of 10.2 (N: 2.2-5.7 mg%) for which we have no explanation.

The patient was discharged to the care of her primary physician and has done well during the subsequent 10 mo of follow-up.

At the time of follow-up, a tomographic whole-body scan was obtained three days after injection of gallium-67 citrate. It showed increased activity, in keeping with the bone-scan findings reported the previous year (Fig. 3).

Trephine bone-marrow biopsy and a bone-marrow aspirate of the left iliac crest were interpreted as consistent with mastocytosis (Fig. 4). The repeated laboratory work was found essentially unchanged.

The patient had suffered from mastocytosis for at least 22 yr and has been under the care of two dermatologists. During this time she has had at least two skin biopsies; the first suggested mastocytosis, the second was diagnostic of mastocytosis on both light and electron microscopy (Fig. 4).

*Received July 1, 1982; revision accepted Feb. 28, 1983.

For reprints contact: Dr. R. D. Ensslen, Dept. of Nuclear Medicine, Cross Cancer Institute, 11560 University Ave., Edmonton, Alberta, Canada T6G 1Z2.

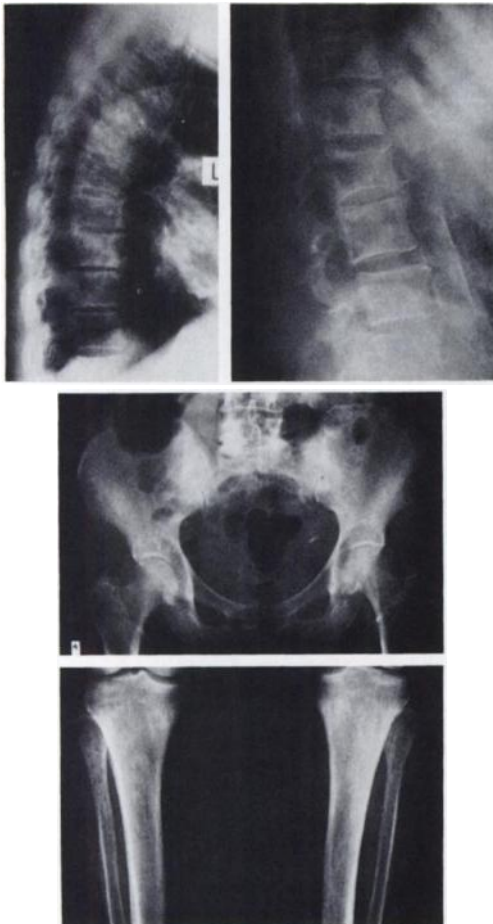


FIG. 1. Radiographs demonstrating sclerosis and thickening of trabeculae compatible with mastocytosis: lateral dorsal spine (upper left), lateral lumbar spine (upper right), pelvis (center), and proximal tibiae and fibulae (bottom).

Reports of radiographs dating back to 1973 do not refer to any significant bony abnormalities, although one dated November 1981 suggested changes as far back as 1976, and the radiographs confirm this.

TABLE 1. SELECTED SKELETAL KILO COUNTS PER MINUTE

Anterior		
1. notch		95 K
2. sternum		74 K
3. xiphoid		44 K
Posterior		
1. cervical		72 K
2. up. dorsal		88 K
3. mid. dorsal		112 K
4. low dorsal		100 K
5. lumbar		95 K
6. sacrum		
left	center	right
115 K	94 K	112 K

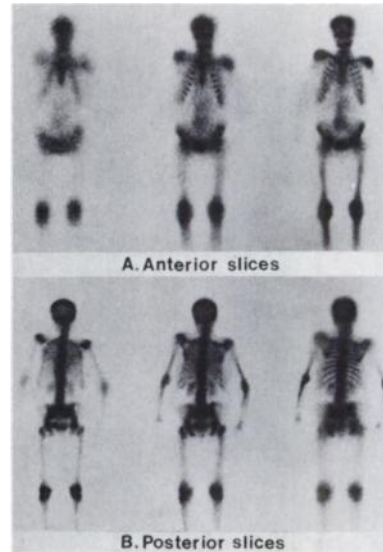


FIG. 2. Tomographic scintigrams* obtained with Tc-99m MDP. These blurred tomographic cuts show increased bony activity throughout skeleton. Kidneys are well visualized.

DISCUSSION

Mastocytosis (urticaria pigmentosa) is a rare disease, usually seen by dermatologists. It is a chronic disorder, considered benign. Its cause is unknown. The diagnosis is confirmed by a properly obtained and stained (toluidine blue) biopsy demonstrating an excessive number of mast cells.

Systemic (2,3) radiographic (1,5-9), and scintigraphic (10-12) findings have been described in the literature.

This patient's findings are characteristic for the disorder. The count rates we obtained on the bone scan clearly showed variable activity in various portions of the skeleton, which has not been described previously. In addition, the kidneys are adequately visualized, unlike in some previous reports (10,11), but in keeping with another (12). The Ga-67 scan is also in keeping with the one previously reported in reflecting the increased bone uptake. The

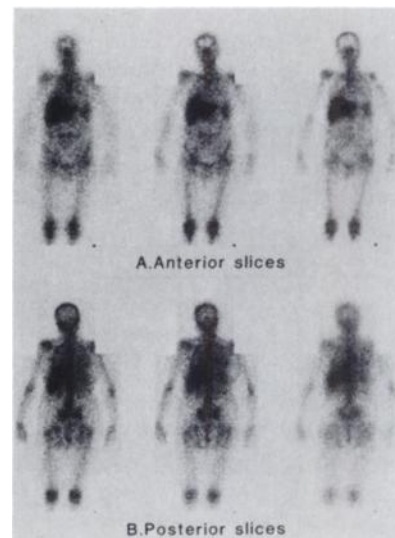


FIG. 3. Tomographic scintigrams obtained with same scanner at day 3 after Ga-67 citrate. The scan simply reflects increased bony activity.

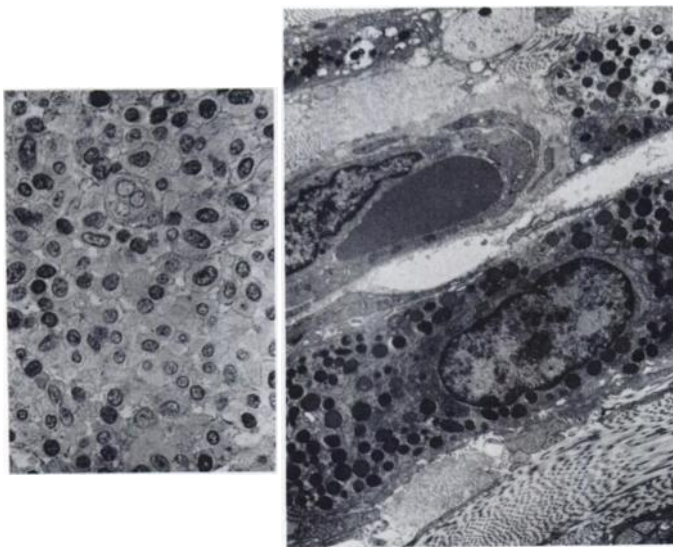


FIG. 4. Photomicrograph showing infiltration of bone marrow by microscopic focus of mast cells (left). Note presence of moderate nuclear pleomorphism, small nucleoli, and altered nucleus-to-cytoplasm ratio in some cells, consistent with neoplasia. X 650, Trephine bone-marrow biopsy fixed in B-5 fixative, embedded in Epon 812-Araldite 502 resin and stained with H&E. Electron micrograph of skin lesion from this patient, showing characteristic and abundant electron-dense granules within cytoplasm of mast cells (right). Appearance of mast cell from marrow would be similar.

condition, although rare, should be suspected when a bone scan shows increased activity throughout the skeleton in a patient with urticaria pigmentosa.

FOOTNOTES

- * Siemens Pho Con.
- † Technicare Dual Probe rectilinear scanner.

ACKNOWLEDGMENTS

We thank Drs. T. K. Shnitka, B. C. Lentle, and G. O. Bain and Mrs. H. C. Dierich for their help and support, and Mrs. Mary Laschowski and Mr. Karl Liesner for their assistance in preparing the manuscript.

REFERENCES

1. CLYMAN SG, REIN CR: Urticaria pigmentosa associated with bone lesions: A survey and report of 8 cases. *J Invest Dermatol* 19:179-185, 1952
2. ENDE N, CHERNISS EI: Splenic mastocytosis. *Blood* 13: 631-641, 1958
3. HOWARD CWH, SCOTT RB: Urticaria pigmentosa with visceral and skeletal lesions. *Q J Med* 28:459-470, 1959
4. NICKEL WR: Urticaria pigmentosa; mastocytosis: a consideration of various manifestations. *Arch Dermatol* 76: 476-498, 1957
5. SAGHER F, COHEN C, SCHORR S: Concomitant bone changes in urticaria pigmentosa. *J Invest Dermatol* 18: 425-432, 1952
6. GAGNON JH, KALZ F, KADRI AM, et al: Mastocytosis: Unusual manifestations; clinical and radiologic changes. *Can Med Assoc J* 112:1329-1332, 1975
7. BENDEL WL, RACE GJ: Urticaria pigmentosa with bone involvement. *J Bone Joint Surg* 45:1043-1056, 1963
8. MACDONALD FR, PEIRCE CB: Urticaria pigmentosa with bone lesions. *J Can Assoc Radiol* 8:15-18, 1957
9. POPPEL MH, GRUBER WF, SILBER R, et al: The roentgen manifestations of urticaria pigmentosa (mastocytosis). *Am J Roentgenol* 82:239-249, 1959
10. SY WM, BONVENTRE MV, CAMERA A: Bone scan in mastocytosis: Case report. *J Nucl Med* 17:699-701, 1976
11. SOSTRE S, HANDLER HL: Bony lesions in systemic mastocytosis. *Arch Dermatol* 113:1245-1247, 1977
12. DERAMOND H, REMOND A, GRUMBACH Y, et al: Bone locations of systemic mastocytosis: radiographic and scintigraphic study (author's translation). *J Radiol (French)*, 61. Aug-Sept. 503-508, 1980
13. CONSTABLE AR, CRANAGE RW: Recognition of the superscan in prostatic bone scintigraphy. *Br J Radiol* 54: 122-125, 1981
14. CONSTABLE AR, CRANAGE RW: Re: Pitfalls of absent or faint kidney sign on bone scan. *J Nucl Med* 22:658, 1981
15. SY WM, PATEL D, FAUNCE H: Significance of absent or faint kidney sign on bone scan. *J Nucl Med* 16:454-456, 1975
16. WITHERSPOON LR, BLONDE L, SHULER SE, et al: Bone scan patterns of patients with diffuse metastatic carcinoma of the axial skeleton. *J Nucl Med* 17:253-257, 1976
17. LUNIA SL, HERAVI M, GOEL V, et al: Pitfalls of absent or faint kidney sign on bone scan. *J Nucl Med* 21:894-895, 1980
18. LUNIA SL, HERAVI M, GOEL V, et al: Re: Pitfalls of absent or faint kidney sign on bone scan. *J Nucl Med* 22:658, 1981
19. SAGHER F, EVEN-PAZ Z: Mastocytosis and the mast cell. Chicago, *Year Book Medicine*, 1967, p 97

Erratum

In the Book Review entitled "Introductory Physics of Nuclear Medicine," Volume 24, p.454, 1983, the review should be attributed to William Dunn, Mayo Clinic, Rochester, Minnesota.