PRELIMINARY AND SHORT REPORT

BONE LESIONS IN URTICARIA PIGMENTOSA

Report of a Central Registry on Skeletal X-Ray Survey*

F. Sagher, M.D. and S. Schorr, M.D.

Following the first observation of generalized osteosclerotic bone lesions in urticaria pigmentosa in 1952 (16), the question arose as to whether this finding is of an accidental nature or whether these bone changes were caused by the same tissue mast cells as the skin lesions. Clyman and Rein (7) also found bone lesions in 2 cases of urticaria pigmentosa, but the roentgenological findings in their cases were of a localized type and different from the original description. In order to find out the types of bone lesions and their incidence, these authors suggested that reports of X-ray findings in urticaria pigmentosa be referred to us as a central registry.

In table I a survey of all cases of urticaria pigmentosa is given in whom X-ray examinations were performed and reported to us since 1952. Cases which were published or presented and were therefore available are included.

The table is presented in chronologic order of the observations reported. In this survey of 52 patients with urticaria pigmentosa, bone lesions were detected in 19 cases. The bone lesions were of two entirely different forms.

The generalized type: Generalized cystic osteoporosis of the ribs, with a thickening of the bony trabeculae; stippling of the bony structure in the skull and thickening of the skull tables; generalized sclerosis of the pelvic bone and vertebrae. A bony pattern consistent predominantly with osteosclerosis. Generalized bone lesions in mast cell disorders different from urticaria pigmentosa were further observed by Degos (10) and Efrati (11) (personal communications). These cases are not included in the table.

The localized type: Calcified deposits and decalcified areas of various size in humerus, radius, femur, skull and shoulder were described by various authors.

The sex of the patients recorded revealed no special preponderance (9 females, 7 males, in 3 not stated). All 4 patients with urticaria pigmentosa and generalized bone lesions were adults, and both patients with mast cell disorders other than U.P. were adults. Among the 15 patients with localized bony changes, 3 were children of 3½ and 5½ years of age and one was 13.

The nature of the bone lesions in one of our patients with generalized osteosclerosis was ascertained by a post-mortem examination in which mast cell aggregates were detected in the affected bones.

CONCLUSION

A report of a central registry of reports of X-ray examinations of bones in urticaria pigmentosa since 1952 is given.

Fifty two patients with urticaria pigmentosa were examined by X-rays and bone changes were detected in 19.

The bone lesions were of two different forms, generalized and localized.

The generalized type was observed so far only in four adult cases; the localized type in adults and children.

These findings indicate that the tissue mast cell has affinity for the bone marrow causing osteoporosis and osteosclerosis.

---

* From the Department of Dermatology and Venereology (Dr. A. Dostrovsky, Director) and the Department of Roentgenology (Dr. A. Druckmann, Director), Hadassah University Hospital, Jerusalem, Israel.

Received for publication March 2, 1956.

TABLE I

X-ray findings in bones in patients with urticaria pigmentosa

Author	No. of Cases Examined	No. of Cases with Positive Findings	Age	Sex		Type of Lesions
				Male	Female	
Sagher, F., Cohen, C. and Schorr, S.: J. Invest. Dermat. **18:** 425, 1952 and unpublished data	15	2	53	1		Sclerotic changes of ribs, pelvis, lumbar vertebrae and small islets of condensation throughout skull.
			55		1	Sclerosed trabeculae of ribs, pelvis, dorsal, lumbar and cervical spine. Numerous tiny islets of increased density throughout skull. Some areas of sclerosed medullary cavities of the extremities.
Clyman, S. G. and Rein, C. R.: J. Invest. Dermat. **19:** 179, 1952 and personal communication	10	2	3		1	Mottled calcified deposits in the upper third of the left and right humerus, upper third of both femurs and slight slcerosis of medullary cavities of the upper third of both tibias.
			40		1	Multiple heredit. osteochondromas.
Calnan, C. D.: Brit. J. Dermat. **65:** 375, 1953 and personal communication	2	2	24		1	Skull: Annular clear cut deficiency about 1 cm diameter of both tables in the left parieto-frontal region in the line of a suture
			52		1	Skull: A small ill-defined defect in the right frontal bone about 5 mm diameter.
Asboe-Hansen, G.: Acta. dermat-venereol. **33:** 471, 1953 and personal communication	1	1	13	1		The epiphysal centre of the left capitulum humeri and both obecrana and left trochlea extremely irregularly divided into several small fragments.
Degos, R.: Actas dermo-sif. **46:** 759, 1955	15	6 (only 3 of them described)	35		1	A few small calcifications not well defined in the femur and skull and the distal part of the left radius.
Grupper, C.: Arch. Dermat. & Syph. **69:** 108, 1954 and personal communication			5		1	Two decalcified areas at the distal part of the right femur near the metaphysis
Parmantier, J.: Thèse, Paris, 1954			21		1	Several areas of decalcification with well defined borders at the right fronto-parietal part of the skull.

Kierland, R. R.: personal communication	2	2	48 47	1	 1	Tiny area of questionable radiolucency in the medial malleolus of the right ankle. Small oval defect in the middle of the right femur.
Bluefarb, S. M. and Salk, M. R.: Arch. Dermat. & Syph. **70:** 376, 1954	1	1	30	1		Generalized cystic osteoporosis of ribs with some coarsening of trabeculae and stippling more pronounced on the medial side of clavicles and similar changes in thoracic and lumbar vertebrae. Mottled osteoslcerosis in the pelvic bones and the upper ends of the femurs.
Reilly, E. B., Shintani, J. and Goodman, J.: Arch. Dermat. & Syph. **71:** 561, 1955 and personal communication	1	1	34	1		Cystic patchy areas of osteoporosis, pagetoid thickening of the bone trabeculae in skull, pelvis, ribs and humeri besides islands resembling bone infarcts in pelvis and humeri.
Cordero, A.: Minerva Dermat. **29:** 55, 1954 abstr. Ann. de dermat. et syph. **82:** 189, 1955	1	1	unknown	1		Osteoporosis of the skull.
Shair, H. M. and Casper, S. L.: personal communication 1955	1	1	5½ months	1		Both radial shafts exhibit an abnormal appearance manifested by irregularity in contour and an absence of the normal cortex which is alternately thinner as well as somewhat thicker than normal.

REFERENCES

1. Asboe-Hansen, G.: Urticaria pigmentosa with bone lesions. Acta dermat.-venereol., **33:** 471–475, 1953.
2. Becker, S. W.: Urticaria pigmentosa. Arch. Dermat. & Syph., **69:** 760, 1954.
3. Berlin, Ch.: Urticaria pigmentosa as a systemic disease. Arch. Dermat. & Syph., **71:** 703–712, 1955.
4. Bertellotti, L.: L'urticaria pigmentosa come reticoloendoteliosi sistemica ad orientamento monomorfo. / A proposito di un caso./ Giorn. ital. di dermat. e sif., **84:** 6891, 1943.
5. Bluefarb, S. M. and Salk, M. R.: Urticaria pigmentosa with bone lesions, gastrointestinal symptoms and splenomegaly. Arch. Dermat. & Syph., **70:** 376–378, 1954.
6. Calnan, C. D.: Urticaria pigmentosa with bone lesions. Two cases. Presented at the Royal Society of Medicine, Section of Dermatol., Brit. J. Dermat., **65:** 375–376, 1953.
7. Clyman, S. G. and Rein, C. R.: Urticaria pigmentosa associated with bone lesions. A survey and report of eight cases. J. Invest. Dermat., **19:** 179–185, 1952.
8. Cochrane, T.: Personal communication.
9. Cordero, A.: Richerche clinico-sperimentali in un caso di "urticaria pigmentosa". Minerva Dermat., **29:** 55, 1954; abstr. Ann. de dermat. et syph., **82:** 189, 1955.
10. Degos, R.: Urticaria pigmentaria y otros tipos de mastocitosis. /Ensayo de Clasificacion de las mastocitosis cutaneas./ Actas dermo-sif., **46:** 759–785, 1955.
11. Efrati, P.: Personal communication.
12. Grupper, C.: Discussion of paper by Sobel, Nathan. Urticaria pigmentosa. Arch. Dermat. & Syph., **69:** 108, 1954.
13. Kierland, R. R.: Personal communication.
14. Parmantier, J.: Les lésions osseuses de l'urticaire pigmentaire. Thèse, Paris, 1954.
15. Reilly, E. B., Shintani, J. and Goodman, J.: Systemic mast-cell disease with urticaria pigmentosa. Arch. Dermat. & Syph., **71:** 561–569, 1955.
16. Sagher, F., Cohen, C. and Schorr, S.: Concomitant bone changes in urticaria pigmentosa. J. Invest. Dermat., **18:** 425, 1952.
17. Shair, H. M. and Casper, S. L.: Personal communication, 1955.