

## REPRINTED ARTICLES

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### LEPROSY: A ROENTGENOLOGICAL SURVEY<sup>1</sup>

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In our study more than a thousand roentgenograms were taken of the patients at Kalihi Hospital, Honolulu, whether or not there was clinical evidence suggestive of bone pathology. All hands and feet were examined, and a sufficient number of the skulls, spines, and long bones to acquaint us with the frequency with which these bones were affected. Serial roentgenograms were made under the same conditions at intervals of about four months on all children under 18 years of age, as it was found that the hands and feet of this group showed the most rapid and marked changes. Roentgenograms were repeated on patients in all age groups who showed evidence of disease of the bones or joints.

The objects of this survey were to learn the bones most commonly involved in the disease, the nature of the involvement, and

<sup>1</sup>This is a condensation of an article which appeared in *The American Journal of Roentgenology and Radium Therapy*, 28 (1932) 598, and which it is felt should be brought widely to the attention of leprologists. It has been provided by the senior author, with the permission of the Surgeon General, United States Public Health Service. The original article is profusely illustrated with reproductions of roentgenograms which cannot be reproduced here; that article should be consulted by anyone especially interested in the subject.—EDITOR.

if possible to establish more definite roentgenological criteria of leprosy. It was also desired to know the frequency and time interval with which bone changes occurred, and whether they had any direct relation to cutaneous and nerve exacerbations.

Of the 140 patients studied—of ages ranging from 6 to 84 years, 64 per cent being from 16 to 40 years—a total of 108 showed some form of roentgen pathology of the bones, only 32 being apparently normal. Changes in the hands and feet were shown by 72, or 50 per cent. The long bones of the arms and legs showed little evidence of disease. In one case there was a cystic degeneration at the distal end of the fibula, and in another at the end of the ulna; a few other long bones showed periostitis and osteitis associated with leprosy infiltration and ulceration of the overlying skin. In a few instances hyperplasia of one or more ribs was seen. The spines were practically negative. In 75 roentgenograms of the skull only one showed bone pathology, namely, destruction in the front bone probably due to syphilis. Fifty cases were studied because of marked soft-tissue involvement of the nose and throat, but in no instance was there found evidence of disease of the nasal bones. This we think is due to the minimal amount of marrow in these bones.

The degree of cutaneous leprosy involvement is not a reliable index to the amount of bone pathology. Bone changes may be independent of or may occur during skin and nerve exacerbations.

#### CLASSIFICATION OF CHANGES FOUND

For descriptive purposes changes found were classified as follows:

A. Those alterations of the bones, particularly of the hands and feet, which showed minute defects such as "nicks," "slices," "frayings," "collar-buttoning," and grossly prominent or enlarged nutrient foramina.

B. Grosser lesions of bones, evidencing osteitis, osteomyelitis, and cystic degeneration ("leprosy osteitis multiplex cystica").

C. Arthritis, destructive and productive.

"Nicks" of class A are small defects such as indentations or excavations of the phalangeal tufts. A part of the tuft may be diagonally shaved off, or "sliced." The tip of the tuft may be combed out, which gives it a frayed appearance, or the entire tuft may have disappeared, giving the remaining phalanx the appearance of a collar button. Enlarged nutrient foramina are small, round,

punched-out areas of rarefaction and absorption at the sites of the normally tiny apertures of entrance of the nutrient vessels. These are rarely seen in normal hands and feet, but their frequency in these cases (about 30 per cent) indicates that they are atrophic responses to a pathological vascular supply.

In the B class, found in 10 per cent of our cases, the changes were seen as increased radiolucency, honeycombing, spotty osteoporosis, eccentric or concentric atrophy, and local lenticular cystic areas with peculiar alterations in the size of structures; there may be an increase or decrease in size of an entire bone. In some cases is seen a "transverse linear atrophy" of the shaft which may progress to a complete solution of continuity, a pathological fracture. This may be the result of nerve impairment, pathological vascular supply, or local infection.

From the foregoing is seen the wide and extensive variety of bone atrophy, decalcification and demineralization which the films disclose in the bones of the hands and feet of lepers. A generalized homogeneous, diffuse atrophy may be the result of, say, ulnar nerve involvement, and may be evident in the last two fingers. Localized, multiple, punched-out areas of decreased density are significant of osteoporosis. In some instances enlarged nutrient foramina could well be pressure or eccentric atrophy, due to perivascular infiltration; this form of atrophy also results from enlarging lepromata. Fraying, nicks and collar-buttoning emphasize concentric atrophy. Slices and transverse linear atrophy are probably the result of neurovascular leprous pathology, though some instances of transverse linear dissolution probably result from local leprous infection.

The softening and disarrangement of bone calcium due to disturbed blood, lymph and nerve supplies to bony centers in young children who have leprosy brings on a sequence of phenomena prominent among which are pain, inflammatory swelling, with increased local heat and tenderness. This condition is exemplified on the film by mushrooming and telescoping of an articular end of a bone of the hand or foot, an osteo-chondritis deformans which is the result of leprosy and in the end leaves an arthritis deformans.

Regeneration or healing of the shafts was noted in many of the phalanges, but destructive processes in the tufts tended never to heal. Transverse linear areas of dissolution in some instances are

replaced by lines of bone condensation. Varying degrees of osteosclerosis replaced areas previously diseased.

We have been led to the belief that the greatest changes in bone leprosy occur during the formative period, that is, at or about the time of union of the integral parts of the bone. It is also observed that rapid improvement in one phalanx may be accompanied by destructive changes in others. It is our belief that in such cases the infection is carried to the bone by way of the vascular supply and there develops into a subacute leprosy osteitis and osteomyelitis. Coincident or subsequent secondary infection by the same route may play a part in the process. The infection once localized in a bone may heal, leaving an area of bone condensation, or it may result in marked destruction with absorption of bone and cystic degeneration. Curettings from such a lesion have been found teeming with acid-fast bacilli. In ten months six patients were seen with acutely swollen painful digits, which we diagnosed as leprosy osteitis.

The patients with arthritis (class C), comprised 27 per cent of our cases. Mechanical arthritides result from relaxation of tendons and joint capsules due to a primary leprosy nerve involvement. Such joints lack the protection afforded by the antagonistic tonus of normal muscles and tendons and become abnormally mobile, and in consequence slight trauma results in destruction of the articulating ends of the bones. Following the primary relaxation there is a secondary contraction, due to destruction of the nerves, that gives the more normal antagonistic muscles and tendons control of the member. Many patients early in the course of their disease show dorsal flexion of the digits and later the characteristic claw hand.

Sensory nerve impairment to pain tends to bone and joint destruction, because injuries are unnoticed until function is disturbed. Fractures unnoticed by patients were discovered in serial roentgenograms. Probably there is arthritis, both inflammatory and toxic in nature, and many joint changes are due to secondary infection by contiguous spread. In the final stage the patients are severely crippled with a deforming arthritis.

#### DIAGNOSIS

When a roentgenogram reveals a combination or association of shadows indicating involvement of the tufts, shafts, and joints of the hand or feet, with changes such as nicks, slices, fraying, collar-buttoning, enlarged nutrient foramina, transverse linear dissolution

near the proximal or distal extremities of the shafts, worm-eaten, or honey-combed appearances of the shafts, leprosy is to be suspected. Further, the phenomena occur in cycles, showing periods of dormancy and reactivation similar to the recessions and exacerbations noted in cutaneous leprosy. These demonstrable alterations in structure are regarded as inflammatory, affecting locally or selectively a bone or bones of the hands and feet. After the initial reaction the bone may regenerate or degenerate. One bone in an extremity may improve at the same time another is retrogressing. This more or less selective, localized sprinkling of pathology in varying stages of degeneration or regeneration is, from a roentgenographic viewpoint, characteristic of leprosy.

Other diseases may give rise to shadows similar to leprosy, notably Raynaud's disease, tuberculosis, syphilis, coccidiosis, scleroderma, so-called sarcoid and other grades of osteomyelitis. Kornblum (4) states that similar changes occur in Raynaud's disease, and that the bone changes of leprosy are not specific. With suspected Raynaud's disease the diagnosis should be in abeyance until leprosy is ruled out. Remote nerve and vascular pathology in leprosy would, in our opinion, more clearly simulate Raynaud's disease than leprosy osteitis and osteomyelitis; these are specific. As regards tuberculosis, the caries and necrosis attributable to leprosy are clear-cut and sharply demarcated on the films, while tuberculous lesions about and near the joints are likely to throw a haze. Slicing or transverse linear atrophy is not likely to occur in tuberculosis. However, it might be impossible to differentiate osteitis tuberculosa multiplex cystica from osteitis leprosa multiplex cystica. Doub and Menagh (2) reported two cases of sarcoid, which in the discussion was considered to be a typical tuberculosis, which could well have been leprosy, the result of guinea pig injections from the infective material being negative. Kohler (3) pictures a rare case in which leprosy could have been the etiological factor. Syphilis and coccidioid granuloma (1) are both likely to show a proliferative periostitis of the hands and feet which on the whole is absent in leprosy. In addition, these diseases tend to affect grossly other bones than those of the hands and feet. Coccidioid granuloma, being rapid in its course, shows less atrophy of adjacent bones. Leprosy does not extend into neighboring joints, and formation of sequestra and involucra is not usually observed in uncomplicated leprosy.

#### CONCLUSIONS

1. Leprous osteitis, osteomyelitis and arthritis occur in bones and joints of the hands and feet, often producing a characteristic appearance in roentgenograms.

2. The bones of the hands and feet of young growing individuals show the most interesting and marked developments, attendant on leprosy infection.

3. Leprous children should have roentgenograms taken of their hands and feet at intervals of three to six months to observe the incidence, variety and evolution of the disease as it affects these extremities. It might be possible to register by this means in suitable cases various effects of different forms of treatment.

4. The most marked changes occur at or about the time of the union of the diaphyses with the epiphyses. Startling improvements have been seen in individual bones in which there was a union of the diaphyses with the epiphyses.

5. Degeneration, regeneration, retrogression and improvement take place in the bones of the hands and feet with about the same constancy as is seen in cutaneous or nerve leprosy (soft tissue leprosy). These processes may be coincidental.

6. Changes for the better or worse in bones of the hands and feet have often been noted going on simultaneously in as short an interval as four to six months. In leprous children, where no previous involvement of bone is discernible, destructive changes attendant on leprous infection may occur within four months or less.

7. A rather striking similarity is noticed in the young, growing bones of the hands and feet, somewhat analogous to that group of juvenile diseases affecting various portions of the skeleton grouped under the category of osteochondritis deformans or osteochondrotropho-dystrophy.

8. Similar and prolonged roentgenological surveys are indicated.

9. Careful evaluation of the roentgenologic findings here enumerated should lead to a suspicion and then a clinching of the diagnosis of leprosy in obscure cases.

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