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MELORHEOSTOSIS

A Case Report with Special Reference to its Mineral Content per Unit Volume

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Melorheostosis is a very rare systemic disease of bone. Its etiology and pathogenesis are still unknown. Since the first case published by Léri & Joanny (1922), about 180 cases including 18 Japanese cases (Araki 1954, Fukuda et al. 1954, Hayashi 1963, Kaneko 1951, Kido et al. 1966, Maenosono et al. 1967, Manabe et al. 1952, Masumi 1963, Munakata et al. 1954, Nakagawa et al. 1952, Sakurabayashi 1941, Sawada et al. 1969, Shinohara 1942, Toriyama et al. 1963, Yamada et al. 1965) of melorheostosis have been reported in the world literature. But its real incidence might be higher than we expect, because some cases have no pain or disabilities. The purpose of this paper is to report an additional case of this condition and to describe the density of bone mineral per unit volume within melorheostotic bone of this case.

CASE REPORT

M. K., a fifty-three year-old Japanese male, was first examined on 27 December 1966, because of a ten-year history of swellings and deformities in his left hand. There was no history of injuries. The diagnosis of melorheostosis was made from its characteristic roentgen appearance. He was admitted to the Nihon University Hospital on 7 January 1967 for further examination and treatment.

Physical examination revealed swellings and deformities on the radial aspect of the left middle finger just distal to the proximal interphalangeal joint and on the dorsum of the left hand corresponding to the third metacarpal shaft. These swellings were not tender to palpation. Any movement of the hand caused no pain in the swollen regions. There were no abnormalities of the soft tissues of the hand.

X-rays revealed the irregular areas of markedly increased density in the proximal and middle phalanges of the middle finger, the second and third meta-

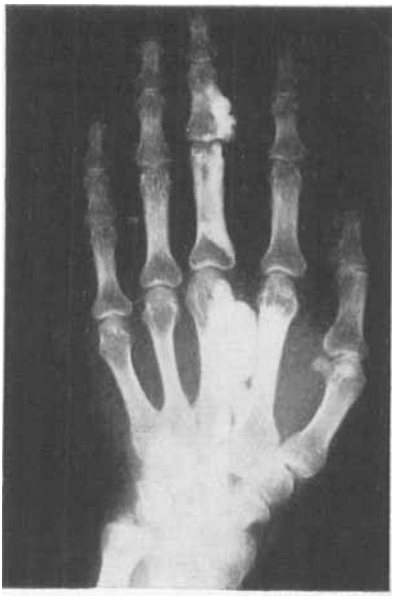

Figure 1. Roentgenogram of the left hand showing the areas of markedly increased radiodensity in the proximal and middle phalanges of the left middle finger, the second and third metacarpal bones, the capitate and lunate bones and the distal end of the ulna.

carpal bones, the capitate and lunate bones and the distal end of the ulna (Figure 1). Cortex was markedly thickened and outline of the middle phalanx and the third metacarpal bone was distorted. Other sclerosed bones were all endosteal or cortical.

Laboratory studies, including complete blood count, urinalysis, plasma alkaline phosphatase, serum calcium, and phosphorus and serum proteins, were all within normal limits.

Resection of the thickened cortices of the middle phalanx and the third metacarpal bone was performed on 11 January 1967. Grossly, the periosteum was normal in appearance, and the cortical bone appeared something like a stalactite.

Microscopic examination of the protruding mass revealed fibrosis in the surrounding connective tissue and active membranous ossification along the margin of the bone. The pattern of the Haversian systems in the underlying bone tissue was irregular in most areas (Figure 2). When examined under the polarizing microscope, the bone tissue consisted mainly of fibrous bone, and matrices around the vascular spaces were replaced by lamellar bone. There were no islands of cartilage with evidence of endochondral ossification and no inflammatory vascular change.

COMMENT

Pain, stiffness, swelling, and deformity are the frequent symptoms and signs, but only characteristic X-rays establish the diagnosis. There are continuous or interrupted streaks or blotches of sclerotic bone along a part or the whole of a tubular bone, suggestive of the flow of wax down a lighted candle. Diagnosis in a typical case is easy, but,

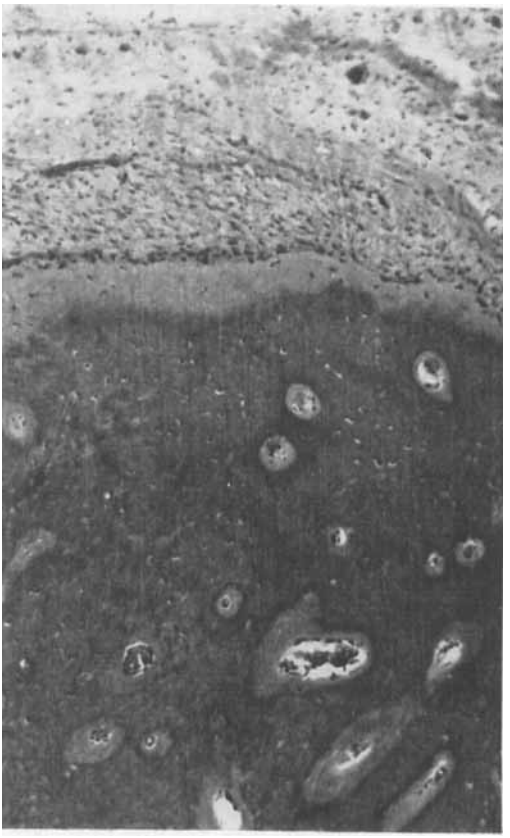

Figure 2. Photomicrograph showing fibrosis in the surrounding connective tissue, membranous ossification and irregular pattern of Haversian systems. (Hematoxylin and eosin, × 130).

as Walker (1964) states, difficult cases are occasionally encountered. He reported two cases of mixed sclerosing bone dystrophies which showed characteristics of osteopoikilosis, osteopathia striata, and melorheostosis.

According to Morris et al. (1963), Ernsting (1966) and Campbell et al. (1968), increased or decreased length, deformities, such as club foot and genu varum, scleroderma, lymphedema, and hemangioma are often associated with this condition. Involvement of the epiphyseal cartilage by osseous lesion is thought to be responsible for occurrence of these limb deformities. Muller et al. (1963) reported seven cases of melorheostosis accompanied by linear scleroderma and suggested that linear scleroderma may represent a primary mesenchymal defect that occasionally spreads into the skeletal tissues. There is no satisfactory explanation for the remaining soft tissue abnormalities.

Biopsy findings in our cases are consistent with these reports except islands of cartilage with the evidence of endochondral ossification. The absence of cartilage formation and endochondral ossification may be due to the situation that the biopsy specimens were from the diaphyseal lesions (Campbell et al. 1968). Furthermore, we could not find inflammatory vascular change in our specimens, which was suggested by Morris et al. (1963).

MICRORADIOGRAPHIC INVESTIGATIONS

In 1946 Engström described a quantitative method for determining the amount of calcium salts in bone using microradiographic techniques. Since that time, refinements in the technique and their theory (Engström 1949, 1962, Lindström 1955, Hoh et al. 1959) and extension of its field of application have provided valuable information about the distribution of various substances, especially hydroxyapatite (Wallgren 1957, Holmstrand 1957, Nilsonne 1959, Ericsson 1965), in microscopic level.

To register the quantitative microradiograms, an X-ray unit of high stability (Philips PW1010), equipped with a copper target, was run at 20 KV and 16 mA. The resultant radiation was then filtered through a 0.02 mm nickel filter. Under these conditions, relatively good monochromatisation was obtained, and it was estimated by Wallgren (1957) and Holmstrand (1957) that more than 80 per cent of the radiation which passed through the nickel lay in the wave length range 1.5 to 1.6 Å, the major part being derived from CuKα radiation.

A thin ground section of biopsy specimen with a step wedge of aluminium foils on Kodak Spectroscopic Plate 649, was exposed to this relatively monochromatic CuKα radiation. After developing the plate, the microdensitometry was performed. Comparing the microphotometric readings of the image of the specimen and the corresponding steps in the wedge, the amounts of hydroxyapatite per unit area can be calculated. In order to know the amounts of hydroxyapatite per unit volume, it is necessary to measure the thickness of the section. In this study the section was cut with a microdissection knife so that all of the points to be measured came to lie at the free cut edge. The thickness of cut edges was measured by micrometry in a profile microscope employing reflected light.

Figure 3 shows two different part of the same microradiogram of

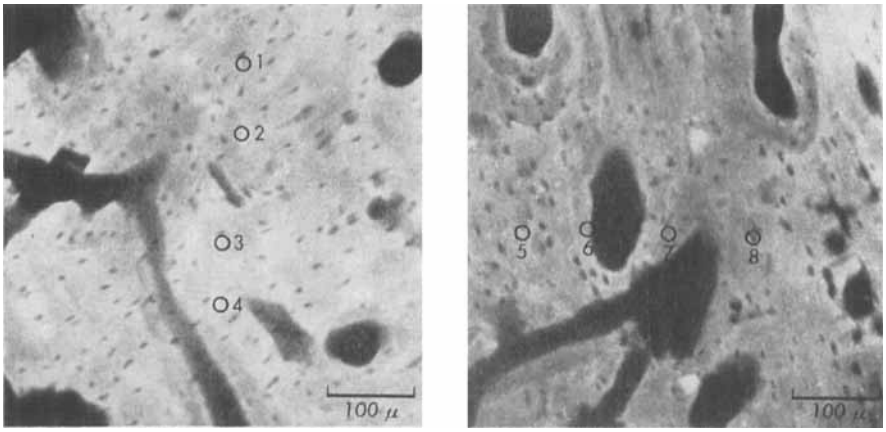

Figure 3. Two different areas of the same microradiogram. The observed density of the specimen at the indicated points is as follows:

Points	Density (gm hydroxyapatite per cm^3)
1	1.05
2	1.09
3	1.07
4	1.12
5	0.93
6	1.02
7	0.94
8	0.94

the specimen. The observed density of the specimen at the indicated points is tabulated. These are the representative points among the forty points measured. The density (grams of hydroxyapatite per cubic centimeter) of the many points of this specimen lay between 1.12 and 0.93 $g.cm^{-3}$. The density of pure hydroxyapatite is 3.15 $g.cm^{-3}$ (Rowland 1959) if calculated on the basis of the unit cell dimension given by Carlström (1955). Then the percentage by volume of hydroxyapatite of the measured points of our specimen lay between 36 and 30 per cent. These values of the specimen are never higher than those of normal human bones.

Gross changes in mineralization are usually diagnosed by visual inspection of the clinical radiogram, but we must always keep in mind that the clinical radiogram is a superimposed image of innumerable structures within the bone, and that visual inspection of black and white in a radiogram would deceive us so that we would have a tremendous difference in mineral content per unit volume. The micro-

scopic density measured revealed that the mineral content per unit volume of the melorheosteotic bone is not higher than that in the normal bone.

SUMMARY

An additional case of melorheostosis is reported. The mineral content per unit volume of the melorheostotic bone of this case was within normal limits.

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