Primary Generalized Hyperostosis in Ancient Peru

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Primary generalized hyperostosis is a rare disease usually diagnosed by radiographic examination. Uehlinger considers this syndrome to be different from the secondary osteoarthropathy of the Marie-Bamberger variety and Paget's disease. The primary type is probably of familial origin and may involve all the bones of the body, appearing independently of severe pulmonary disease. It is predominantly a disease of males (29:2) and in some cases is associated with pachydermia.

The literature on this disease is primarily European; the case described below is the first in South America and first among pre-Columbian people.

Materials and Methods. Four bones consisting of a left femur, a right hip bone, and two ribs from a single skeleton were recovered from the surface of a Huari culture cemetery (about 1000 A.D.) near El Ingenio in the Ingenio Valley, Ica, Peru. Grave robbers had desecrated the cemetery and opened the mummy bundles looking for valuables. These bones were collected by one of us (Sotil) and taken to the Regional Museum of Ica for study.

X-rays were taken of all bones, and one rib was used for histological study. Several sections were cut on a band saw and ground to a thickness of 80µ to 100µ.

Results. The hip bone (os coxis) was that of a female estimated to be 32 years old by utilizing the pubic symphysis technique of Gilbert. This bone weighed 480 gm compared with 180 gm for a normal bone of this size female of a similar age. The left femur was 405 mm long, 50 mm in diameter (midshaft) and weighed 750 gm (Fig 1). The marrow cavity measured 8 mm to 15 mm by x-ray (Fig 2). A normal femur from a woman this size and age had a

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Fig 1—The femur from the present case of primary generalized hyperostosis is compared to a normal femur from a woman of the same size. The first weighed 750 gm while the latter weighed 350 gm.

Fig 2—Radiograph of the diseased femur shows some reduction in size of marrow cavity with tremendous thickening of the shaft. Articular surfaces are essentially normal. Rib radiographs showed complete obliteration of marrow cavity.

weight of 350 gm with a diameter of 30 mm; the marrow cavity measured 9 mm to 28 mm. The two ribs each weighed 110 gm compared to 30 gm for a normal rib. All control bones were also from the Huari culture.

Histologically the major finding was an increase in the circumferential lamellae as a series of periosteal proliferations occurring at different times and giving the multilayer effect seen in Figure 3. An increase in Volkmann’s canals was also noted. The endosteum had also been active as the marrow cavity virtually disappeared with randomly arranged new haversian systems and interstitial lamellae as seen in Figure 4. In certain areas, as seen in this figure, occasional erythrocytes and hemoglobin pigment may be noted as well as calcified remains of blood vessel walls.

Conclusion. The bones described above lack the soft tissue for study, but it is probable that this individual belonged to the group with pachydermia. This observation is based on the extensive disease seen in the femur which is more compatible with hyperostosis associated with pachydermia. Hyperostosis without the skin lesions is usually located on the distal end of the diaphysis.

Although a rare disease, this case is of interest, for the diagnosis is generally a radiological one. Since it is thought to be of familial origin, it is important to document its geographic distribution in the world. This case is the first reported from pre-Columbian South America and in an American Indian.
Fig 3—Low power view of the circumferential lamellae of an 80µ ground section of rib shows a series of periosteal proliferations. Numerous Volkmann’s canals were seen in other sections (×60).

Fig 4—View of the area of the marrow cavity revealed replacement of the cavity with randomly arranged new haversian systems and interstitial lamellae. Occasional erythrocytes (arrow) in 80µ ground section of rib (×100).

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