CALCINOSIS

Report of Two Cases

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In broad terms, the classification of calcification of soft parts is divided into four groups: metastatic, dystrophic, myositis ossificans and calcinosi; but, as the pathogenesis of many of the "calcifications" is unknown, such a classification must be an arbitrary one, and not always satisfactory.

Thus in the group of metastatic calcifications there are some that are associated with a wide variety of other conditions such as nephritis, multiple myelomatosis, myeloid leukaemia, sarcoma, hyperparathyroidism, Albers-Schönberg disease, and even osteomyelitis and caries of the spine. Changes in calcium metabolism are demonstrable in only a few of these conditions—notably in hyperparathyroidism.

In the dystrophic calcification group are included, for example, hydatid cysts and valvular and tendon calcifications. A separate group is formed for the myositis ossificans of traumatic origin, and with these should be included the paracordial ossification of the Pellegrini-Stieda type, ossifications of the coraco-clavicular ligaments and other calcareous deposits of clearly traumatic origin.

Calcinosi, on the other hand, is an anatomical, pathological, radiological and clinical entity showing pathological calcification in which multiple deposits of calcium are laid down in the skin, in the subcutaneous tissues and in connecting structures of deeper tissues; so far no case has been described in which calcification in internal organs or viscera has been demonstrated, either on radiographic examination or at necropsy.

The first case of calcinosi was described by Teissier in 1877. Steinitz (1931) reviewed the literature of calcinosi and divided it into two groups: "calcinosi circumscripta" and "calcinosi universalis." As the pathogenesis of the affection is still obscure, however, the division of calcinosi into two groups is a somewhat forced one, seeing that not even the topographic distribution of lesions allows a sufficiently distinct dividing line to be drawn always between the two alleged forms. Despite this limitation we must mention that "calcinosi circumscripta"—also described as calcareous granuloma, subcutaneous calculus, subcutaneous lithopexia, and calcareous gout—occurs mostly in adults and more frequently in women (in a ratio of six to one), the calcareous deposits appearing in the upper limbs, especially in the fingers. In "calcinosi universalis" the calcareous deposits occur not only in the superficial but also in the deeper tissues and show a tendency to spread, which is slow and intermittent. It is somewhat more frequent in the very young. The "universal" type is less common than the "circumscribed."

In the published discussions on the etiology of this curious pathological process, some of the opinions expressed appear to have been dictated by the preferences of their authors or have been due to more or less circumstantial evidence. Generally speaking, those inclined to the "inner" view look upon the disease ab initio and consider that the observed deposition of calcareous salts represents more than a form of fixation of the calcium in tissues previously altered by causes still unknown. Other authors seek a direct cause-effect relation, linking the calcareous deposits with biochemical and endocrine disturbances not yet understood or demonstrable. Others continue to inquire into the possibilities of a common etiology with apparently allied affections such as scleroderma and sclerodactyly (Thibierge-Weissenbach
syndrome), Raynaud's disease, acrocyanosis, fibrous myositis, hyperparathyroidism and hypervitaminosis D; the latter two parallelisms have been derived from animal tests.

The diagnosis of calcinosis rests upon the demonstration of the calcium deposits of characteristic type and distribution. It has to be distinguished from calcified lipomas, phleboliths, calcified ganglia, metastatic calcifications, gout, myositis ossificans and dystrophic calcifications.

Calcinosis is compatible with reasonable longevity, though function of the locomotor system is often seriously impaired and cosmetic blemishes may be severe. Spontaneous cure has sometimes been observed. No effective treatment is known.
CASE REPORTS

Case 1. Calcinosi s universalis with congenital dislocation of the hip—Girl aged three years at first attendance. At the age of two years a limp had been noticed and radiographs showed a congenital dislocation of the right hip (Fig. 1). There were no signs of abnormal deposits in the soft tissues of the thighs. For reasons not known she received no treatment for the hip. In July 1950 she was referred to this hospital where further radiographs confirmed the existence of a congenital dislocation of the hip, but also revealed the existence of calcinosi s (Fig. 2). There were numerous areas of calcification, some rounded, others in thick bars, grouped around the iliac bones and the proximal epiphyses of the femora. In the right thigh there was a thick bar due to the confluence of small oval elements of high density. Further medially there was another similar bar, extending obliquely to the junction of the middle and lowest thirds of the thigh in the line of the muscle planes. In the left thigh areas of calcification were apparent on the medial aspect only. Radiographic examination of the rest of the body did not reveal other deposits. Blood examination showed no significant abnormality.

Progress—Attempted manipulative reduction of the dislocation was unsuccessful and operation was advised. The parents were slow in accepting this advice so that the operation was not performed until the child was four years old. Nine months after the operation radiographs showed that not only had reduction been maintained but also that the calcinosi s appeared to be completely arrested. Further examination another year later showed no extension of the lesions (Fig. 3).

Comment—The curious aspect of this case is the demonstration of a pathological process which produced extensive calcareous deposits in less than a year and yet became stabilised, with no further extension of the lesions, within fourteen months of the first radiological confirmation.

Case 2. Calcinosi s universalis—Man aged forty-six years. Radiographic examination undertaken on account of pain in the spine and hips revealed evidence of calcinosi s in both thighs (Fig. 4). Radiographs of the rest of the body were normal. Examination of the blood showed normal findings.

Comment—The calcinosi s in this case was discovered incidentally in the course of routine examination for back pain.

REFERENCES

