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CALCINOSIS

(Demonstration of a case)

Generally distinction is made between two forms of pathological calcification within the organism: the dystrophic and the metastatic. Exemples of the former group are found in the areas of calcification in tissues undergoing necrosis, chronic inflammatory processes or senile changes; examples of the latter group are found in the metastatic depots of lime salts under parathyroid hyperfunction, renal rickets, etc.

Calcinosis is a disease we are not yet able with certainty to classify as belonging to either of these two groups, because we do not know anything about its pathogenesis. Calcinosis may be regarded most properly as a tendency of the connective tissue for some unknown reason to retain lime salts. The disease appears partly in a benign circumscribed form, partly in a malignant universal form.

Circumscribed calcinosis, improperly designated as "lime gout", is a relatively common morbid condition in middle-aged and elderly people. It often is combined with Raynaud's disease and sclerodermia. It has a marked tendency to localize to the peripheral parts of the extremities. Generally it causes but little inconvenience.

Universal calcinosis is considerably more infrequent, only thirty odd cases having been described in the literature. The disease occurs in younger persons, and often it starts in early childhood. The case I shall present here belongs to this group of calcinosis.

The typical feature of universal calcinosis is depositing of

larger or smaller amounts of lime (calcium phosphate, calcium carbonate) in the connective tissue anywhere in the body, but mostly subcutaneously, in the interstices between muscles and in fasciæ, besides in the perineural connective tissue, but never in the internal organs or in the connective tissue surrounding these organs. In most of these cases the depositing of lime salts is particularly localized to the surroundings of the large joints (*i.e.*, the shoulder and pectoralis regions, the hip, gluteal and knee regions). It is interesting that, according to Wells, the proportion between Ca, PO_4 and CO_2 in the lime depots is always the same as that between the same substances in bones —namely, 1: 5.7: 0.8, respectively.

The lime depots keep getting larger, often in periodical attacks. Sometimes greater or smaller amounts of these lime salts are discharged, with signs of inflammatory processes: the skin breaks and fragments of lime are evacuated, not infrequently in the form of a cream-like suspension of lime salts mixed with pus. Even up to 2 liters of lime masses have been seen to pour out from a focus at one time, as reported, for instance, by Neuwirth, Krausse and Trappe. In connection with this discharge, as a rule, the nearest joints are swollen and show also other signs of inflammation, often associated with a high rise in temperature. Gradually there comes a temporary improvement and the sores through which the lime has been discharged usually heal well, leaving scars in which, strange to say, usually no new depot of lime appears. But the skin and subcutis of these areas undergo shrinkage.

On account of the increase in volume, these lime depots become more and more troublesome, and the joints become immobilized, one after the other. Paralyses and pareses develop, due to involvement of the perineural connective tissue. Towards the end a cachexia-like condition develops, with a marked degree of muscular atrophy and emaciation. The bones become atrophic from inactivity, but they show no abnormal processes due to disturbances in their lime content.

All attempts to interpret this disease as due to some metabolic disturbances have turned out negative. The calcium and

mineral metabolism has been the subject of particular thorough studies, among others by Teisser, Weil, Weber, and Friedländer. Except in a couple of cases, all blood examinations have given perfectly normal findings.

Attempts to explain the disease as an endocrine disturbance have failed too. Several of these cases have been combined with endocrine disturbances, but nothing has been found as pointing towards any certain organ in this respect. Theories have been advanced that try to interpret the disease as due to some inflammatory process. Other theories have tried to connect calcinosis with rheumatic complaints.

In the Scandinavian countries, Professor Salvesen, Oslo, has paid particular attention to this disease. He has described 2 cases, one in Acta med. Scandinav. 1937 (vol. 92), the other in Nordisk Medicinsk Tidskrift 1938 (No. 13, March), where he has given a thorough account of his metabolism determinations and tolerance tests on these patients. Also his findings show that the calcinosis patients react like perfectly normal persons in calcium tolerance tests, blood examinations, metabolic experiments, alkali reserve determinations, etc.

One of these normal properties is, that when the organism is supplied with a diet low in calcium content, it excretes more calcium than it receives. When the organism is acidified by administration of ammonium chloride, the negative balance concerning the calcium metabolism is increased—and this is a normal condition too.

Salvesen made the interesting observation, however, that the calcinosis patients made up for the deficient calcium supply by taking calcium from the large depots. Under observation for a considerable length of time the depots were thus decreased in a surprising degree. So in this way he obtained a therapeutic effect through the low calcium supply + administration of ammonium chloride.

In our patient the first calcium nodules appeared at the age of 4 years. She is now 12 years old and the lime deposits have increased gradually to its present great extent, of which we see an example in the roentgenograms taken of her pelvic region.

Apart from this she presents no signs of any disease, and the values for blood calcium are normal. Since November 17, 1937, she has been under treatment in keeping with the principles given by Professor Salvesen.

Thus she is given a diet with a calcium content of about

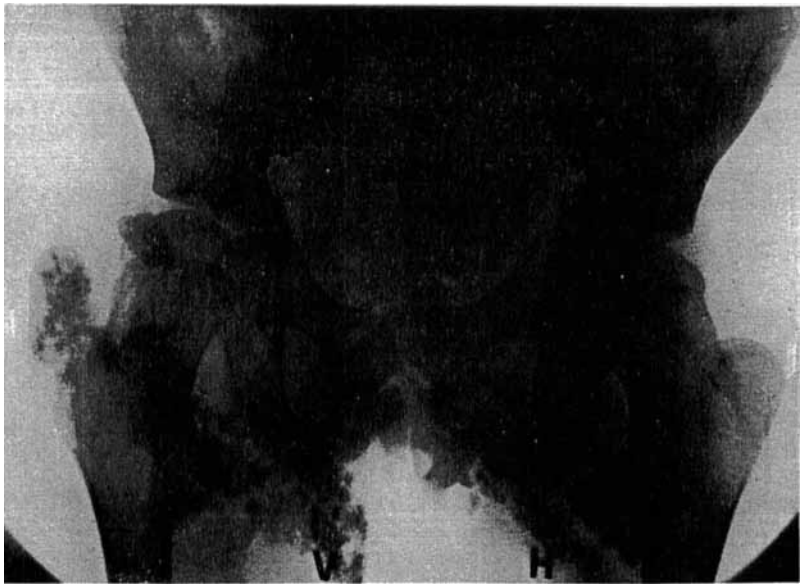

Fig. 1.

124 mg. per day. In addition she is given ammonium chloride, 1 g. 3 times daily, every other week.

Ever since this treatment was instituted she has done all her school work, except in some short periods when she has been hospitalized for the sake of analyses on the excretion of urine and fæces, biopsy of a calcium infiltration, etc.

She has been improving gradually—at any rate distinctly, although not considerably. She feels that now she is moving about more easily, and her mother claims that now she is more bright and lively than before. When lying down, she is now able by herself to get up in sitting posture—something she could not do before.

On physical examination we have been able to observe a distinct diminishing of the depots in the gluteal regions, and a slight decrease in the right pectoralis region. Previously the skin over the gluteal muscles were studded with a considerable number of fungus-like, lime-filled, nodules, varying in size (up

Fig. 2.

to that of a hazel-nut), and now the skin of this region has become plain and smooth. The mobility of the right foot, right elbow and left wrist has increased somewhat. In the gluteal regions, especially on the left side, some lime has been discharged through the skin.

Also by roentgenography is it possible to ascertain a decrease in the calcium depots, although this is distinct only in the pelvic region (see Figs. 1 and 2). No new calcium infiltration has appeared since the institution of treatment. The first roentgenogram was taken on October 5, 1937, the other on August 30, 1938. So the observation period is a little less than 11 months.

The calcium infiltration that is seen at the left greater trochanter on Roentgenogram 1 was extirpated, so this has not disappeared through effect from the internal treatment. Further, as mentioned before, some lime has been discharged through the skin. On comparison of the two roentgenograms, however, it can be seen distinctly that the streaky calcium infiltrations medial to the lesser trochanter have become smaller and less compact.

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In Vol. XI, Fasc. 1. wird ein Generalregister zum Vol. I—X aufgenommen.

Dans Vol. XI, Fasc. 1. va paraitre un table des matières general de Vol. I—X.