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From the Orthopaedic Clinic of the University, Athens, Greece.

# TUMORAL LIPO-CALCINOSIS

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The term tumoral calcinosis was first used by Inclan in 1943 to describe the deposition of dense nodular calcareous masses in the tissues about the hips, shoulders and elbows. Prior to this the term lipocalcinoma-granulomatosis had been employed by Teutschlaender in 1935. For a long time the two above entities have been considered different subgroups of calcinosis (Danowski 1962). Subsequent study of various cases indicated that tumoral calcinosis and lipocalcinoma-granulomatosis were the same conditions (Smit & Schmaman 1967).

## MAIN CHARACTERISTICS OF THE DISEASE

– Tumoral calcinosis occurs in young individuals of both sexes during the first or second decade. There is probably a familial tendency. Local factors such as injury may play a part.

– The exact cause of the disease is still obscure; it has been reported to be a metabolic disturbance of unknown aetiology (Lafferty et al. 1965).

– Certain biochemical findings have suggested that it may be an inborn error of phosphorus metabolism. There is no evidence of hyperparathyroidism, renal disease or excessive intake of vitamin D.

– The course is benign but excision of the masses should be undertaken early to avoid operative difficulties and secondary infection. Chronic multiple fistulae lead to profound cachexia and secondary amyloid disease.

– Histologically, the tumor shows calcific deposits in fibrous stroma with a few inflammatory cells and foreign-body giant cells. Infiltrations of xanthoma cells are present.

– Since tumoral calcinosis is a form of heterotopic calcification, the differential diagnosis includes the following conditions: (a) Dystrophic

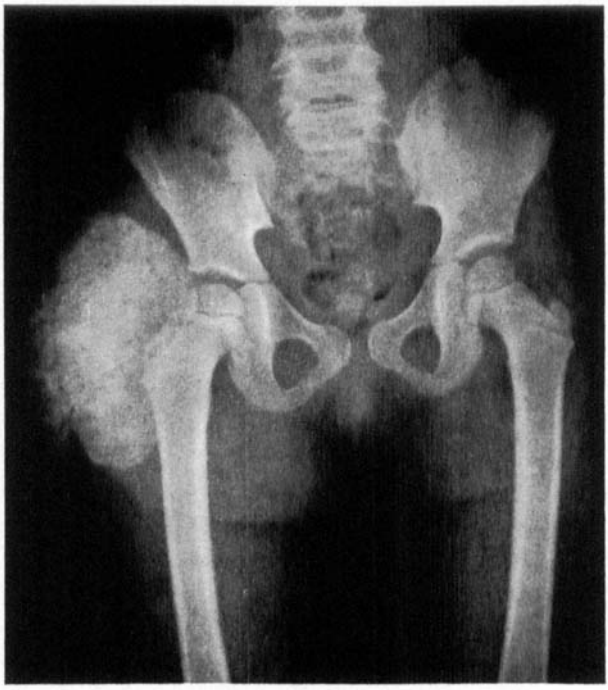

*Figure 1. Radiograph showing multinodular calcaneous deposits around the right hip.*

calcification (dead parasites and fat necrosis); (b) Metastatic calcification in which the usual primary cause is hypercalcaemia. Two forms of metastatic calcification are the hypervitaminosis D and the milk-alkali syndrome; (c) Calcinosis universalis; and (4) Calcinosis circumscripta.

– Treatment is mainly by surgical excision. Prognosis is good although new masses may appear around other joints as the patient ages.

## CASE REPORT

The following case concerns a girl, four years of age, whose clinical picture, laboratory data and microscopic findings coincide exactly with the findings of the so-called tumoral calcinosis.

This female child has a negative family history for this affection. In 1967 a mass was noted in the right hip. It had not caused pain or limitation of motion. The mass was excised and later a similar tumor began to develop in the same region extending forward and backward above the greater trochanter. It had grown steadily, reaching its maximum size in about a year (Figure 1). The recurring tumor was fairly well defined, lobulated, and fluctuating. This tumor had never been accompanied by pain, tenderness, or limitation of motion.

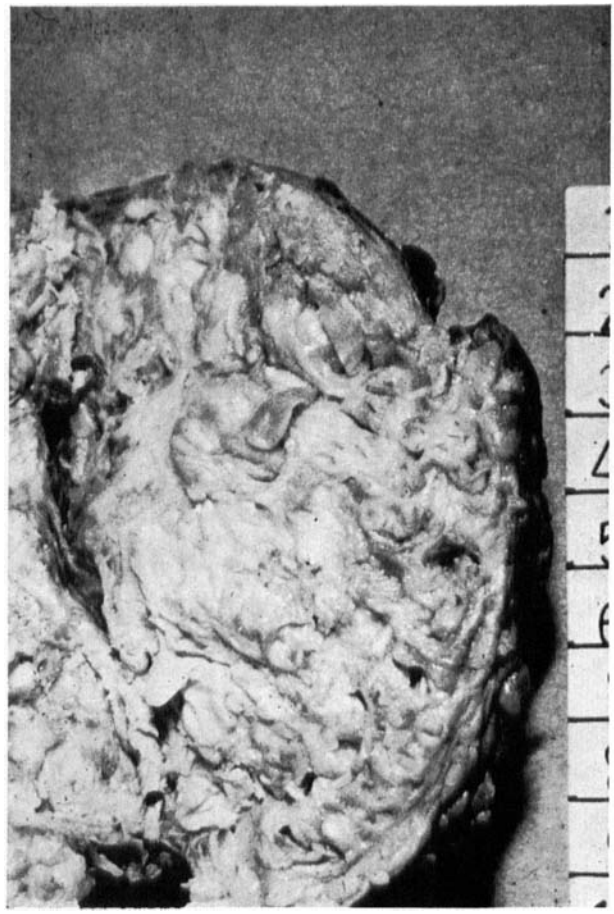

*Figure 2. A photograph of the mass after excision. Cut surface with fibrous stroma and small cavities containing milky fluid.*

Laboratory investigations, including serum calcium and phosphorus and alkaline phosphatase levels, were within normal limits. The second operation revealed a large tumor mass, about 9.5 by 7 by 5.5 centimeters in size. The microscopic examination confirmed the diagnosis.

Histologically, according to the opinion of the pathologist (A. Theodossiou), it seems probable that this rare process presents phases of development: probably, the original lesion consists of deposits of lipoid substances (sudanophil guttulae) into connective cells (Figures 5a and b). The second phase concerns the progressive growth of the lipoid infiltrations. During this phase calcific deposition takes place and a liquidation of the lipoid accumulations appears (Figures 5c and d). Finally, the third phase corresponds to the development, around these spaces, of an absorbing granuloma with mononuclear and polynuclear xanthoma cells (Figures 5e and f). Hence, the term tumoral lipo-calcinosis seems to be more suitable.

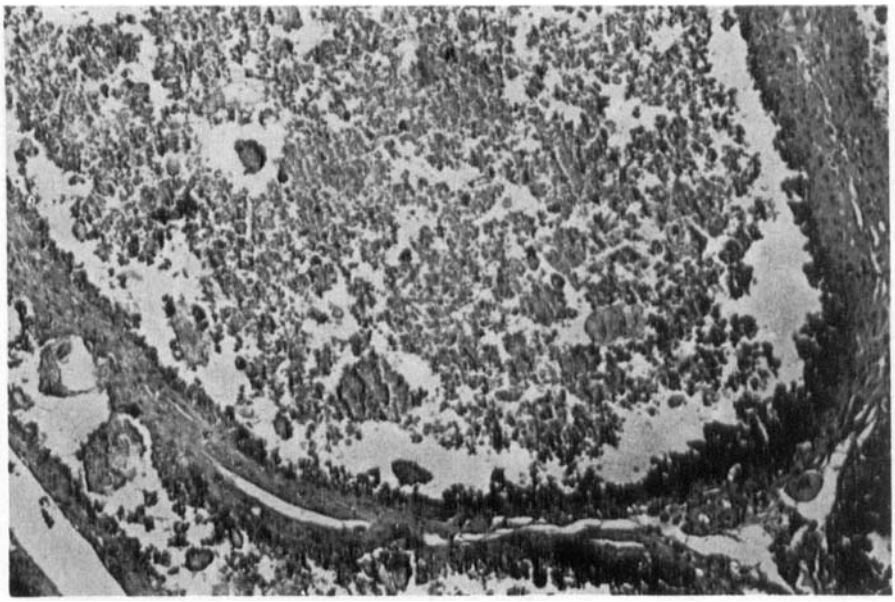

*Figure 3. Photomicrograph stained with Hematoxylin-eosin (× 120). A part of small pseudocyst containing amorphous masses of lipoid and calcium salts.*

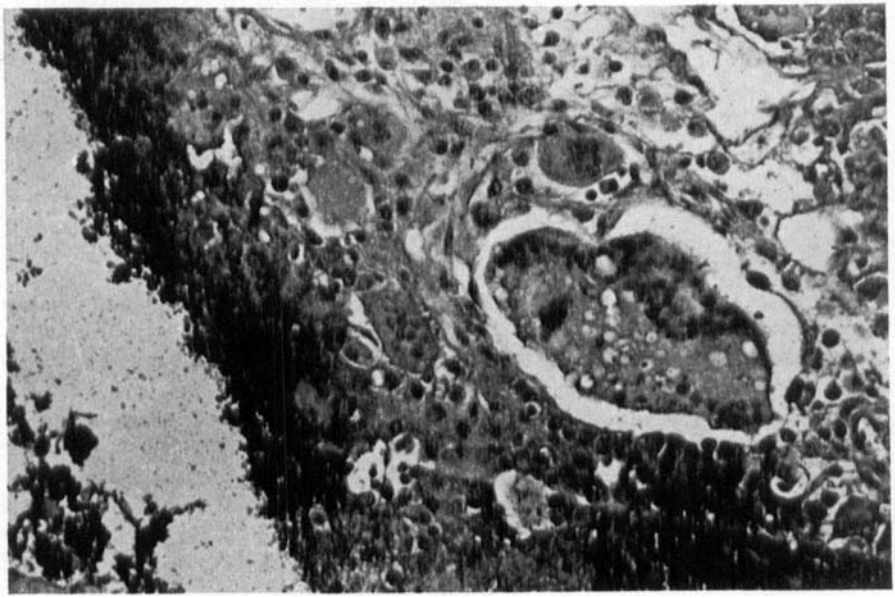

*Figure 4. Another part of the specimen with foreign-body giant cells including guttulae of lipoid substances. Hematoxylin and eosin, × 400.*

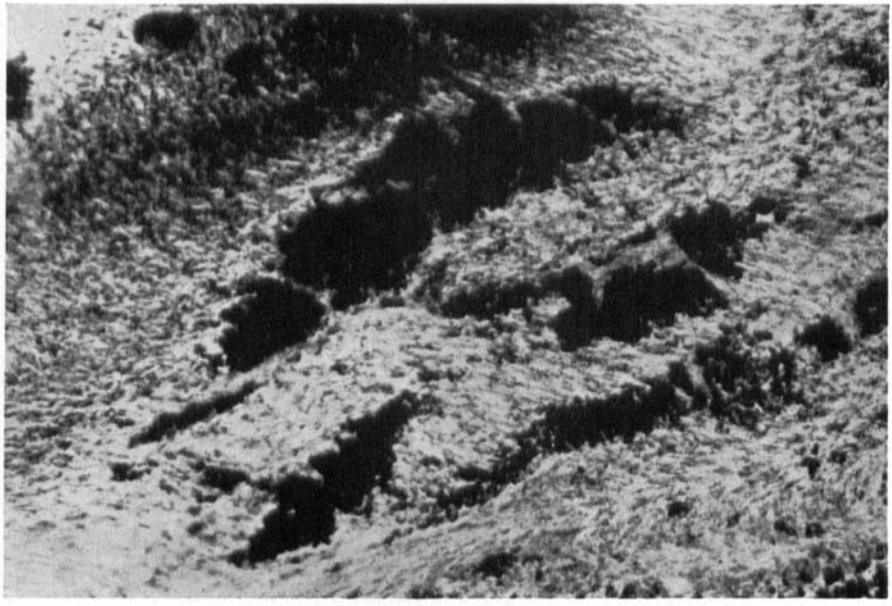

*Figure 5a.*

*Figures 5a, b, c, d, e and f: A series from different sections showing the possible phases of development of the lesion.*

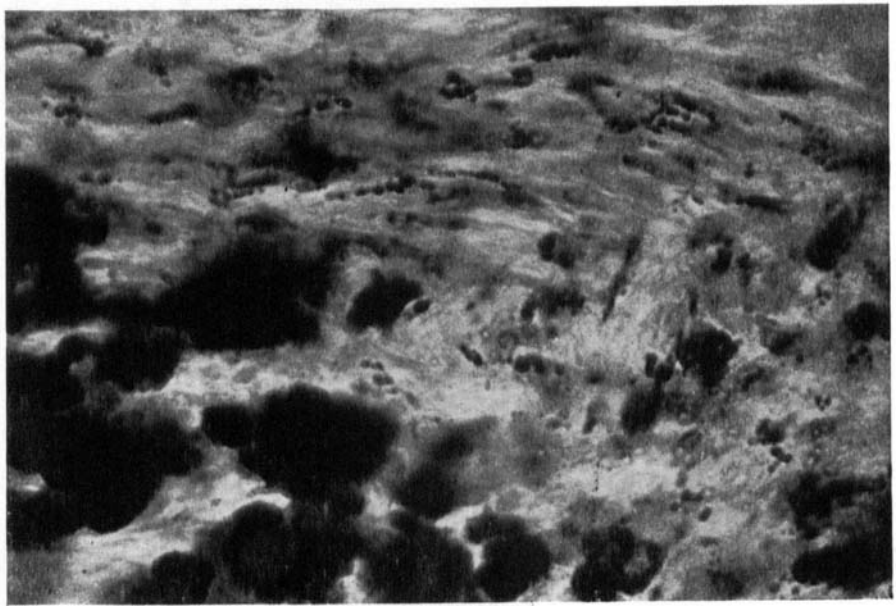

***Figure 5b.***

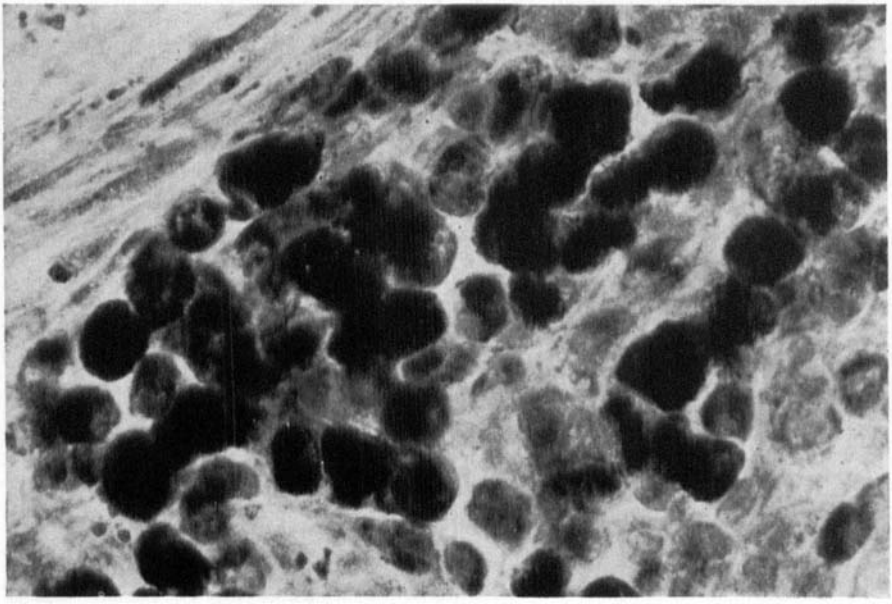

*Figure 5c.*

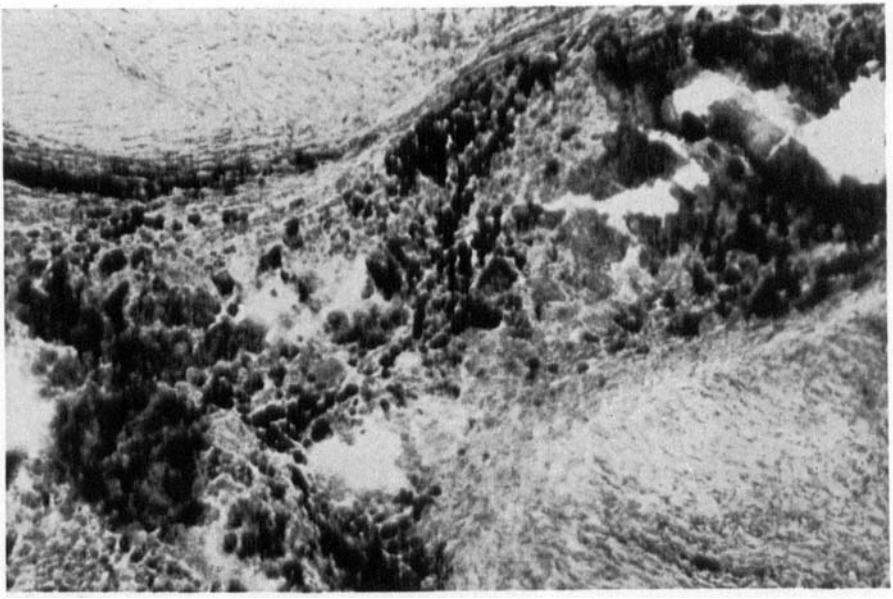

***Figure 5d.***

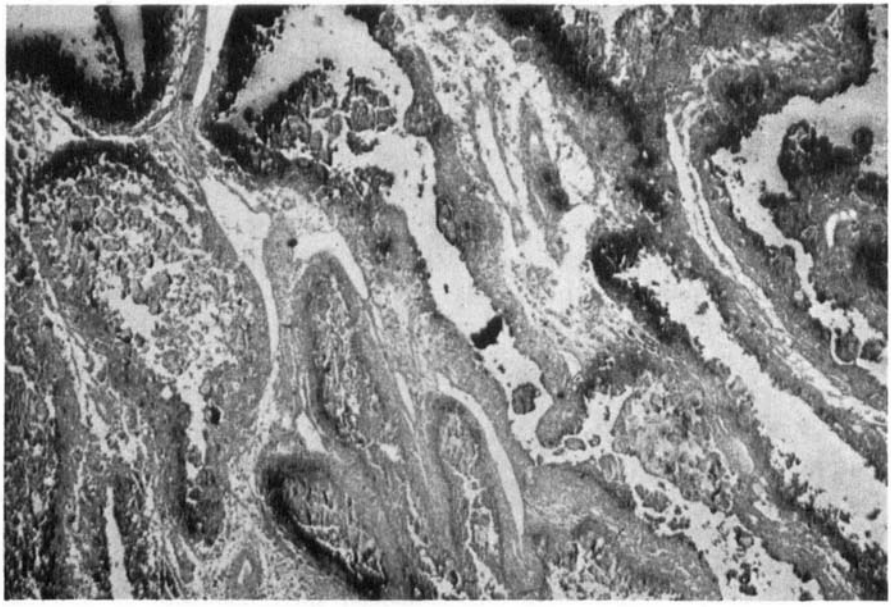

*Figure 5e.*

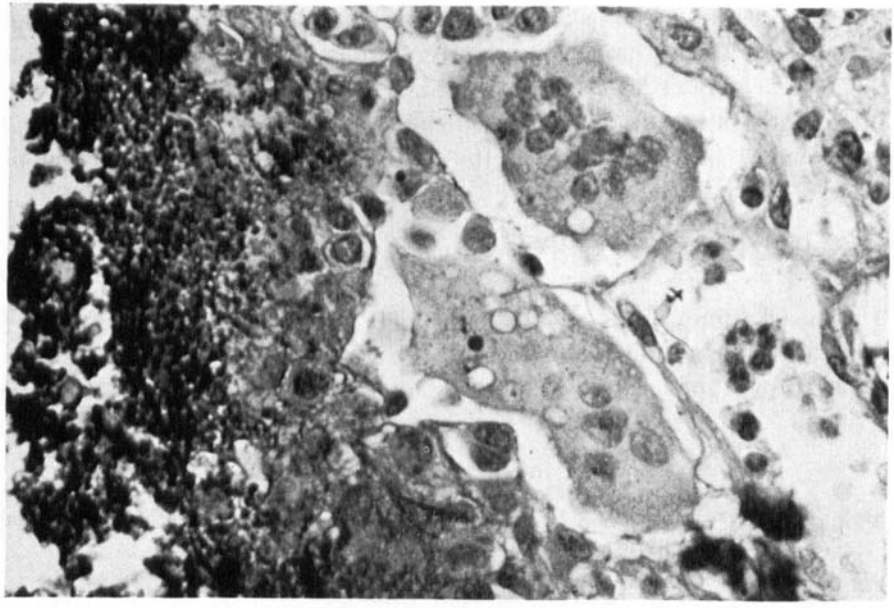

***Figure 5f.***

## COMMENT

Abnormal calcification in soft tissues may assume a variety of forms. The term calcinosis has usually indicated the presence of calcium deposits in extravisceral sites occurring in patients without any discernible abnormality of serum calcium and phosphorus levels. The term metastatic calcification has been employed, in the main, in reference to abnormal deposits of calcium in either visceral or extravisceral sites in patients with disturbances of calcium and phosphorus levels. The origin of calcinosis remains obscure.

Tumoral calcinosis angioid streaks in the retina have also been reported. This finding is associated with two other conditions. These are: pseudoxanthoma elasticum and Paget's disease of bone. Although Paget's disease, pseudoxanthoma elasticum and tumoral calcinosis may be basically distinct entities, the fact that angioid streaks (which are an integral part of a definite disorder of connective tissue) are present, suggests that tumoral calcinosis may likewise be a disorder of connective tissue.

All the above entities are characterized by deposition of calcium salts on the different tissues (McKusick 1961). Nevertheless, all of these probably have a hereditary background.

On the other hand, electron microscopic studies of the lesion in one case of tumoral calcinosis showed that the cells lining the spaces appeared structurally and functionally similar to osteoblasts and osteoclasts (Lafferty et al. 1965).

Is it possible that this form of calcinosis may represent a clinical fragment of an abiotrophy of the connective tissue? Is it a form of extra-osseous Paget's disease?

## SUMMARY

A case of tumoral lipo-calcinosis, in an otherwise healthy girl aged four, is reported. This term seems to be more concordant with the microscopic appearance. The risk of post-operative recurrence is stressed.

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