TUMORAL CALCINOSIS, A CLINICAL REPORT OF ELEVEN CASES

J. F. NOYEZ[1], S. M. MURPHREE[2], K. CHEN[2]

Clinical observations of 11 new cases of tumoral calcinosis are reported. The condition is characterized by calcified masses of varying size in the region of major joints. Surgical excision is recommended in selected cases determined by the size of the lesion, the deformity present and functional complaints. In this series surgical excision was done in 6 patients, and the diagnosis was confirmed histopathologically. After a complete excision of the tumor, no recurrences were seen in 5 cases with a mean follow-up time of 25 months. Incomplete excision led to multiple recurrences in one patient.

Keywords : tumoral calcinosis ; surgical excision ; recurrences.
Mots-clés : calcinose tumorale ; excision chirurgicale ; récidives.

INTRODUCTION

Duret (3) in 1899 described the development of masses of calcium comparable to an orange in size, overlying major joints in a healthy 12-year-old white girl. The name tumoral calcinosis was first given by Inclan (11), who described the syndrome in three unrelated teenage Blacks. Laferty, Reynolds and Pearson (14) in 1966 could only collect 17 cases from the literature and added one of their own. In 1966 Palmer (21) reported on another 50 cases, in addition to the existing 25 cases recorded in the literature.

Three major groups of different clinical conditions with soft tissue calcium deposits can be identified according to their etiopathogenesis : (1) metastatic calcifications, (2) dystrophic calcifications and (3) idiopathic calcifications.

Metastatic calcifications can occur in vitamin D intoxication, milk-alkali syndrome, primary hyperparathyroidism and chronic nephritis due to calcium- and/or phosphorus abnormality. In dystrophic calcifications, calcium is deposited in dead or previously damaged tissue. In idiopathic calcifications there is a complete absence of any other abnormality and this is most commonly represented by tumoral calcinosis. The disease is usually described as a rare condition but probably occurs fairly frequently, although it is seldom recognized in the early stages.

This paper represents the clinical experience of an orthopedic surgeon working in a referral university teaching hospital, reporting on 11 new cases of tumoral calcinosis, seen during a period of only 14 months (July 1989 to August 1990). Five patients were treated surgically, and the diagnosis was confirmed histologically. The other 6 cases were diagnosed as tumoral calcinosis on the basis of the typical clinical and radiological features. The data on age, sex, location, clinical findings and follow-up after surgical excision in 6 patients are reported.

RESULTS

The 11 patients showed characteristic clinical and radiological features of tumoral calcinosis. All

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patients were Black, 8 were male and 3 female. The mean age was 9 years with a range of 6 to 26 years. None of the patients were related. Mean follow-up time was 37 months with a range of 18 to 60 months.

In every case the clinical history was that of a slow growing lump developing over a few years. The most common site was the hip (8 out of 11 patients with 14 hips involved), followed by the elbow (7 out of 11 patients with 11 elbows involved). The shoulder region was affected in 5 patients, the knees in 2, the feet and hands in one patient. In 2 of the 11 patients the lesion was a solitary mass while in 9 there were multiple lesions. In 6 hips, 4 elbows and 2 shoulders the lesions were bilateral.

The tumors usually felt firm. However in 2 cases the consistency was rather soft. In all except one case the overlying skin was intact. In this patient (case 5) the tumors intermittently drained a milk-likefluid. In 7 patients there was functional limitation secondary to the size and location of the tumor. The joints were not involved. Six of the patients had surgical treatment, the main indication being functional deficit. Pain was never a major clinical feature. There was no nerve involvement. One patient (case 5) received successive operations, and multiple recurrences were evident after incomplete surgical excision. In the other 5 cases treated surgically by complete excision, there was no evidence of recurrence after a mean follow-up time of 25 months. No visceral or cutaneous involvement was present.

None of the patients was originally diagnosed as having tumoral calcinosis. One case was referred as a chondrosarcoma, 2 as ectopic calcifictions, one as a calcified hematoma, one as a calcified granuloma, one as cystic calcification and 5 were referred without specific diagnosis. The clinical findings are summarized in table I.

No abnormalities in serum calcium or phosphorus levels were detected in the 5 patients in our series treated surgically.

The roentgenograms were characteristic in all of the patients and showed round or oval masses of varying sizes located around the periarticular soft tissue. The masses appeared lobulated as a conglomeration of multiple round opacities, unattached to bone (fig. 1). Flattening of the posterior aspect of the lower end of the humerus was seen in 2 of the elbows involved (fig. 2).

Table I

case	sex	age	location	original diagnosis	treatment
1	F	7	Hips Elbows	Ectopic calcification	surgery (elbow)
2	M	10	Hip	Chondrosarcoma	surgery
3	M	8	Hips Elbows Shoulder	Ectopic calcification	surgery (hip)
4	M	7	Foot	Calcified hematoma	surgery
5	M	26	Around large joints	—	multiple excisions
6	F	8	Hips Elbows	—	surgery (hip)
7	M	7	Shoulders Elbows	Cystic calcifications	—
8	M	7	Hips	—	—
9	F	6	Shoulders Elbow	—	—
10	M	8	Hips Shoulder	—	—
11	M	6	Hips Elbows	Calcified granuloma	

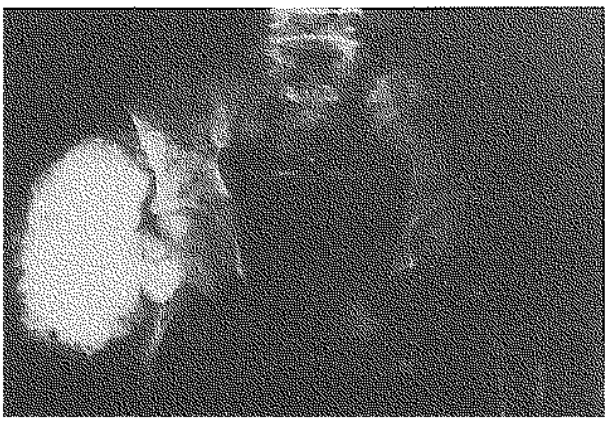

Fig. 1. — Roentgenogram of the pelvis in an 8-year-old boy showing a large mass consisting of a conglomeration of multiple round opacities around the right hip (case 3).

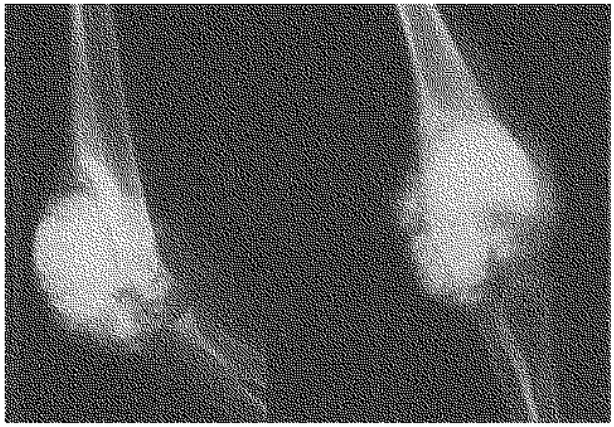

Fig. 2a

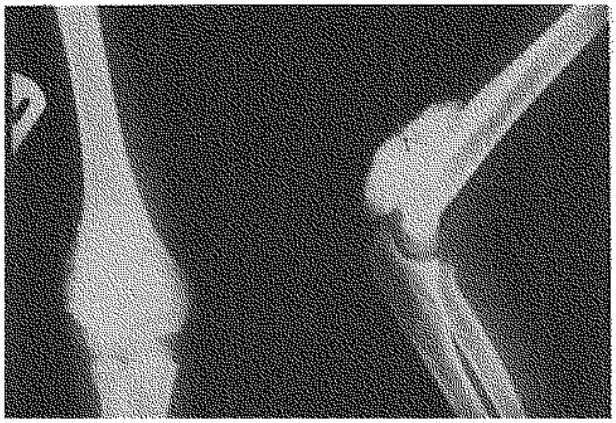

Fig. 2b

Fig. 2. — Roentgenograms of the right (a) and left elbow (b) in a 7-year-old girl showing the typical flattening of the posterior aspect of the lower end of the humerus (case 1).

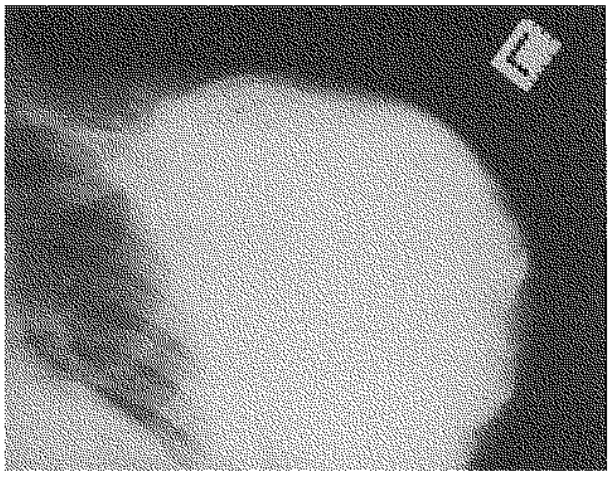

Fig. 3. — Roentgenogram of the left shoulder in an 8-year-old boy showing a huge mass around the entire shoulder girdle (case 10).

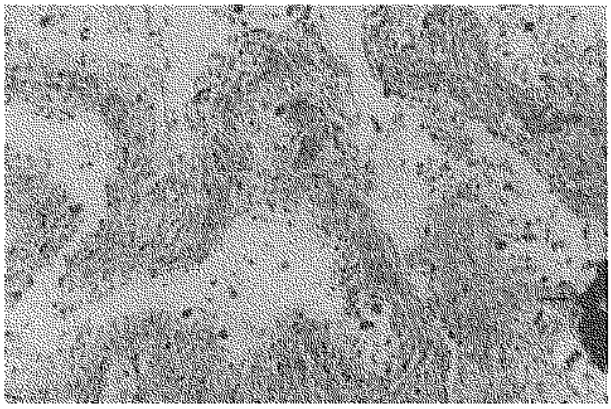

Fig. 4. — Microscopic view showing cystic spaces lined by histiocytes and containing granular calcium. Dense aggregate of calcium present at lower right corner (arrow). (Hematoxylin and eosin, magnification × 110).

Macroscopically the masses were multiloculated and were white to pale-yellow. The sizes of the tumors varied. The largest masses were usually seen around the hip, elbow and shoulders. The largest tumor was present around the shoulder girdle and measured fifteen by twenty-two by fifteen centimeters (fig. 3).

The cases examined histologically, showed the typical features recognized as tumoral calcinosis : microcrystalline, aggregated and granular calcium lying in masses surrounded by thin fibrous tissue septa (fig. 4).

Within lining septa and partly engulfing calcium deposits were macrophages and multinucleated foreign body type giant cells (fig. 5). Inflammation formed a minor component, but granulation tissue merging with fibrous tissue of septa was seen.

DISCUSSION

Tumoral calcinosis is a type of idiopathic calcinosis, characterized by periarticular calcified masses of varying size in the region of major joints

(fig. 6). The hip and elbow are the commonest locations, followed by the shoulders and less frequently the hands, feet and knees. This was confirmed by this study, as shown in table I.

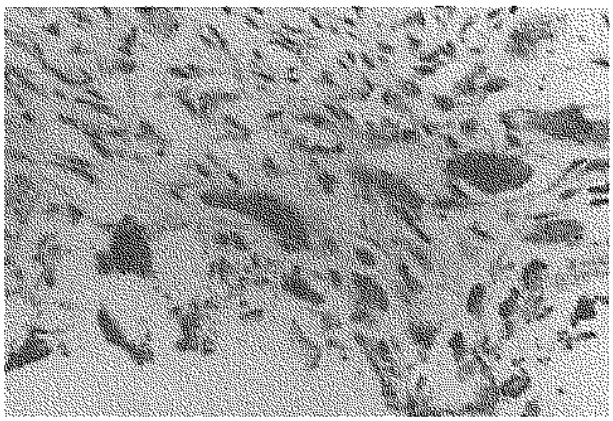

Fig. 5. — Lining of cyst showing granulation tissue and multinucleated giant cells. Fine granular and coarse-calcium lies on the surface. (Hematoxylin and eosin stain, magnification × 470).

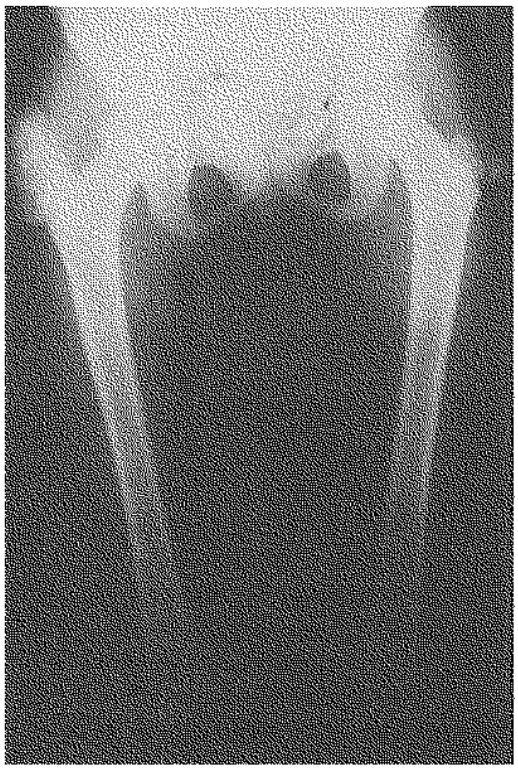

Fig. 6. — Roentgenogram of the pelvis in a 7-year-old girl (case 1) showing bilateral periarticular calcified masses of varying sizes.

The extensor aspect of a large joint is the usual site of the lesion with frequent limitation of function. When the tumor is growing the patient may present with swelling and functional limitation. Usually there is no pain except when there is pressure on the nerves. Most patients in this study who presented with a tumor of large size had some functional limitation, mainly lack of extension of the elbow. Only one patient in this series had severe complaints of pain (case 5). This patient presented with large draining masses around both shoulders after repeat surgery.

On examination the tumor usually feels firm. Macroscopically these tumors are multilocular and cystic, containing a creamy white fluid (fig. 7). They can drain spontaneously, but even in the absence of drainage the masses can fluctuate in size. No visceral or muscular involvement is reported, and this was confirmed by this study.

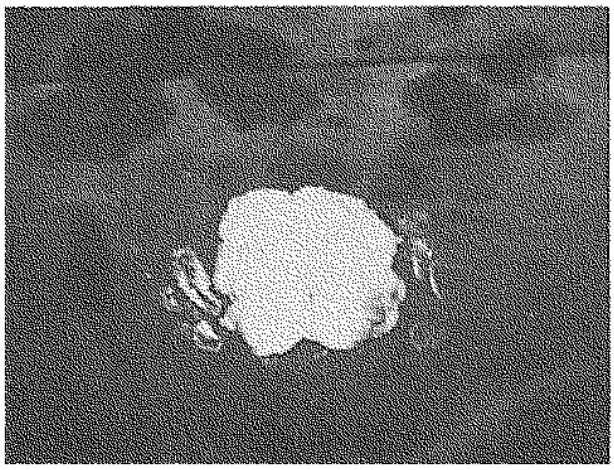

Fig. 7. — Macroscopic view of a resected mass of tumoral calcinosis, showing the creamy white fluid on cross-section.

The majority of cases occur in male, most commonly black patients, and the condition is more frequently observed in the second and third decades of life. However there are cases reported ranging in age from 10 days to 83 years, although only 4 cases in the literature were reported in patients younger than 2 years old (1, 2, 22). In our study, 10 (91%) patients were younger than 11 years of age, the mean age being 9 with a range

of 6 to 26 years. McKee *et al.* (17) reported a 2 to 1 female to male ratio, but most of the authors find a male predominance. This was observed in our series, which included 8 male and 3 female patients.

Histologically, microcrystalline, aggregated and granular calcium masses surrounded by thin fibrous tissue septa are seen. Cystic spaces are lined by histiocytes and multinucleated foreign body type giant cells containing granular calcium. Inflammation forms a minor component, but granulomatous or dense collagenous tissue merging with fibrous tissue is seen.

Laboratory examinations show no abnormality of calcium levels, but hyperphosphatemia is reported in some cases (4, 14). In the 5 patients treated surgically in our series no abnormalities in calcium or phosphorus could be detected.

Radiological examination was as described in other reports on tumoral calcinosis in the literature, showing round or oval masses or calcifications located around the periarticular soft tissue. These masses appeared lobulated as a conglomeration of multiple round opacities, unattached to bone. At the elbow, flattening of the posterior aspect of the lower end of the humerus is a common finding in the presence of a large mass. This radiological feature was present in 2 (18%) of the 11 elbows seen in this study.

The cause of tumoral calcinosis remains obscure, although it was studied by many authors (5, 7, 9, 14, 16, 18, 23). The natural course of the disease is one of a slow progressive growth of the tumors resulting in occasional pain, swelling and functional limitation.

Six patients in our series underwent surgery determined by the extent of the tumor and functional limitation. Full surgical removal is essential, and if removal is incomplete, recurrence is frequent. Administration of steroids, diphosphonates, phenylbutazone, calcitonin and radiation therapy as described in earlier reports (10, 12, 14) had little or no effect. In view of the frequent failure of surgery reported in the literature and the growth of recurrent masses frequently more rapidly than the initial lesion, the results of medical treatment are noteworthy (6, 12, 13, 14, 19, 20).

In our series no such medical management was initiated. A prospective study, evaluating the effect of a pharmacological treatment on a larger number of patients, could however be suggested. Surgical excision was attempted only if the size of the tumor and the functional complaints justified surgical intervention. Care was taken to resect the tumor completely, since incomplete resection yields poor results with recurrences of the masses.

In the 5 patients treated with a complete surgical excision no recurrences were evident. The one patient treated by incomplete surgical excision needed successive operations for multiple recurrences.

In conclusion, wide surgical excision was an effective treatment for tumoral calcinosis in this group of patients with a follow-up time of 25 months.

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SAMENVATTING

J. F. NOYEZ, S. M. MURPHREE, K. CHEN. Tumorale calcinosis. Bespreking van 11 gevallen.

Een klinische studie betreffende elf nieuwe gevallen van tumorale calcinosis wordt voorgesteld.
Deze pathologie wordt gekenmerkt door calcifiërende massa's ter hoogte van de grote gewrichten.
Een chirurgische excisie kan aangewezen zijn afhankelijk van het volume van de tumor, de misvorming en de functionele klachten. In onze reeks werden zes patiënten heelkundig behandeld en de diagnosis werd steeds histopathologisch bevestigd.
Na een volledige excisie van de tumor werden geen recidieven vastgesteld bij vijf patiënten met een gemiddelde follow-up tijd van vijfentwintig maand.
Onvolledige excisie bij één patiënt gaf aanleiding tot meervoudige recidieven.

RÉSUMÉ

J. F. NOYEZ, S. M. MURPHREE, K. CHEN. Calcinose tumorale : présentation de 11 cas.

Une étude clinique concernant onze nouveaux cas de calcinosis tumorale est présentée. Cette pathologie se caractérise par des masses calcifiées au niveau des grandes articulations. Une exérèse chirurgicale peut être envisagée. Ceci dépend du volume de la tumeur, de la malformation qui en résulte et des symptômes fonctionnels.
Dans notre série, six patients ont bénéficié d'un traitement chirurgical. Le diagnostic fut à chaque fois confirmé par l'examen anatomopathologique.
Dans cinq cas d'exérèse complète, aucune récidive n'est apparue en un délais moyen de vingt-cinq mois. Une exérèse incomplète s'est soldée par plusieurs récidives.