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Articles

Tumoral calcinosis: radiologic–pathologic correlation

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Abstract. *Objective.* Tumoral calcinosis is a frequently misdiagnosed disorder. This study details the radiologic and pathologic characteristics of tumoral calcinosis that distinguish it from most other entities.
Design. Radiologic and pathologic findings, and medical records of 12 patients with tumoral calcinosis were reviewed and compared with equivalent information about 5 patients with other calcified lesions.
Patients. The 12 patients ranged in age from 15 months to 62 years. Six had idiopathic tumoral calcinosis and 6 had secondary tumoral calcinosis.
Results and conclusions. A consistent radiologic finding for tumoral calcinosis was a dense calcified mass that was homogeneous except for a "chicken wire" pattern of lucencies, which correlated histologically with thin fibrous septae. Other characteristics of tumoral calcinosis included fluid-calcium levels, demonstrated in four patients, and smooth osseous erosions adjacent to the mass, demonstrated in three patients. Five cases of tumoral calcinosis were originally confused with other calcified lesions; however, the radiologic findings were characteristic of tumoral calcinosis in retrospect.

Key words: Tumoral calcinosis – Soft tissue calcification – Periarticular calcification – Metastatic calcification

Tumoral calcinosis is a condition that can be idiopathic, but is more commonly associated with abnormal phosphate metabolism with a high calcium-phosphate product, above 75 mg/100 ml, often due to secondary hyperparathyroidism [1–4]. This entity has a distinctive radiographic appearance, which is often useful in distinguishing it from other lesions. In this study, we reviewed the imaging and pathologic characteristics of tumoral calcinosis and contrasted them with those of other benign and malignant calcified soft tissue masses.

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Materials and methods

We reviewed radiologic findings, medical records, and pathologic findings of 12 patients with tumoral calcinosis (Table 1). Six of the patients had idiopathic tumoral calcinosis and the other 6 had secondary tumoral calcinosis related to chronic renal disease. Of the patients with idiopathic tumoral calcinosis, 3 were male and 3 were female. Two patients were black and 4 were white. These patients ranged in age from 15 months to 62 years. A 9-year-old girl and a 58-year-old man had hyperphosphatemia, which has been sporadically shown in patients with idiopathic tumoral calcinosis.

Of the patients with secondary tumoral calcinosis related to chronic renal disease, all were white men ranging in age from 30 to 72 years. All of these patients were on dialysis and 4 had confirmed hyperphosphatemia ranging from 6.1 to 10.0 mg/dl.

All 12 patients with tumoral calcinosis had undergone conventional radiography. Two of those patients had also had ^{99m}Tc-MDP scans. CT scans had been performed on 5 patients, and MRI scans had been performed on 4 patients, using a 1.5-T unit (General Electric, Milwaukee, Wisc.). Follow-up radiographs had been taken in 2 patients, at 1 and 4 years, respectively.

In addition, we compared the radiologic and pathologic features in five examples of other dystrophic/neoplastic conditions associated with calcification or ossification that could conceivably be mistaken for tumoral calcinosis (Table 2). They were selected from the radiology files because they presented radiographically as large calcified or ossified masses. These cases included osteosarcoma, chondrosarcoma, synovial cell sarcoma, myositis ossificans, and scleroderma.

Results

The most common locations for tumoral calcinosis in the 12 patients were the shoulders and hips, which were involved in 6 and 5 patients respectively. The elbows and knees were involved in 2 patients each. More unusual locations were the ischium, sternum, and forearm, each seen in 1 patient.

A consistent radiologic finding on conventional radiographs and CT was that of a dense calcified mass that was homogeneous except for a "chicken wire" pattern of lucencies within the mass that gave the calcifications a "cobblestone" appearance (Fig. 1A). These lucencies correlated histologically with thin fibrous septa (Fig.

Table 1. Clinical findings and imaging characteristics in patients with tumoral calcinosis

Case no.	Patient			Location	Imaging studies[a]	Imaging characteristics			Blood phosphate level
	Age (years)	Race (B/W)	Sex (M/F)			Chicken wire septae	Calcium-fluid levels	Bone erosion	
Primary tumoral calcinosis									
1.	56	W	F	Hip	BS, MR	+	+	–	
2.	62	B	M	Hip, elbows	CT	+	+	–	Normal
3.	58	W	M	Hip, shoulders		+	–	–	Increased
4.	1	W	M	Knees		+	–	–	
5.	42	W	F	Ischium	CT	+	–	–	
6.	9	B	F	Hip	BS	+	–	–	Increased
Secondary tumoral calcinosis									
7.	39	W	M	Knees, axilla, shoulder	MR	+	–	+	Increased
8.	30	W	M	Sternum	CT	+	–	+	Increased
9.	48	W	M	Scapula, clavicle	MR	+	–	–	Increased
10.	72	W	M	Clavicular fossa	CT, MR	+	–	–	
11.	50	W	M	Forearm, shoulder, elbow		+	+	–	Increased
12.	58	W	M	Shoulders, hip	CT	+	–	+	

[a] In addition to conventional radiographs
B, Black; W, white; BS, ^{99m}Tc-MDP scan

Table 2. Clinical findings and imaging characteristics in representative patients with other calcified masses

Case no	Patient			Diagnosis/location	Imaging studies[a]	Imaging characteristics		
	Age (years)	Race (B/W)	Sex (M/F)			Chicken wire septae	Calcium-fluid levels	Bone erosion
13.	62	W	F	Osteosarcoma/shoulder	MR	–	–	–
14.	55	W	M	Chondrosarcoma/buttock	CT, MR	–	–	–
15.	20	W	F	Synovial sarcoma/shoulder	CT, MR	–	–	–
16.	15	W	M	Myositis ossificans/thigh		–	–	–
17.	37	W	F	Scleroderma/hands		+	–	–

[a] In addition to conventional radiographs
B, black; W, white

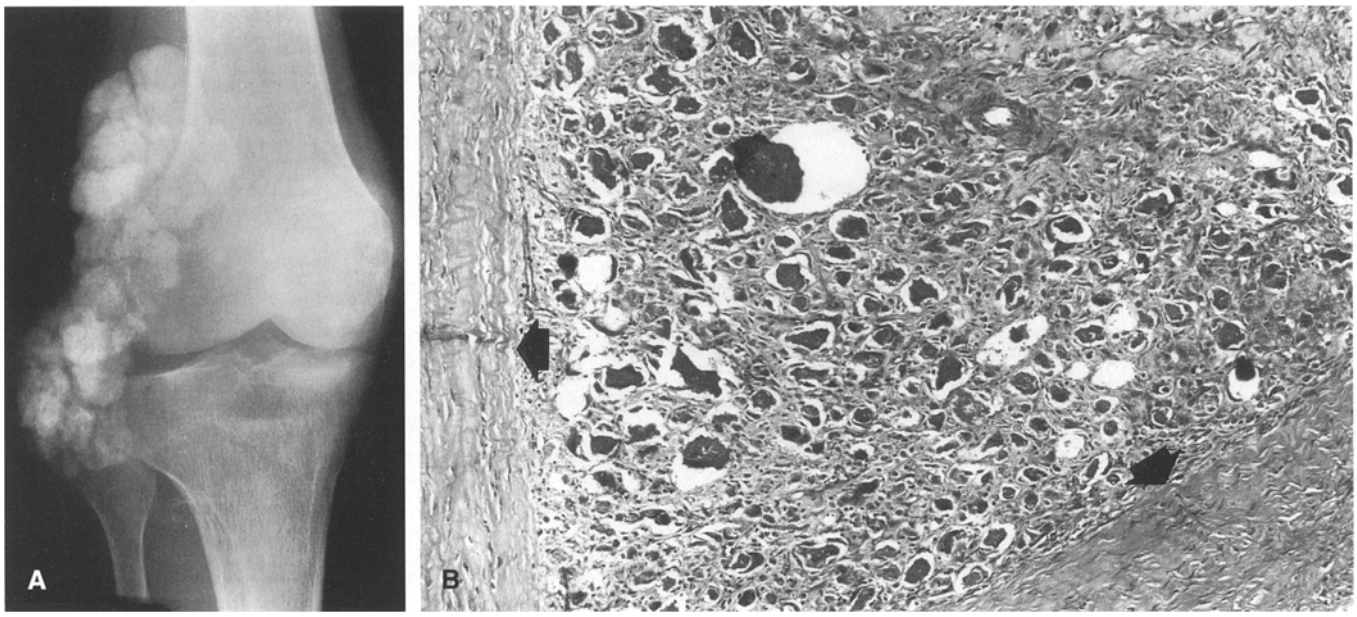


Fig. 1A, B. Case 7. A 39-year-old man who was on renal dialysis for 11 years and was hyperphosphatemic noted a mass in his right knee. **A** The anteroposterior (AP) radiograph reveals homogeneous densely calcified masses separated by low-density septae along the lateral aspect of the knee. This was diagnosed as secondary tumoral calcinosis, and the mass was removed. **B** A histologic section of the tissue shows fibrous bands *(arrows)* surrounding intralobular calcific debris

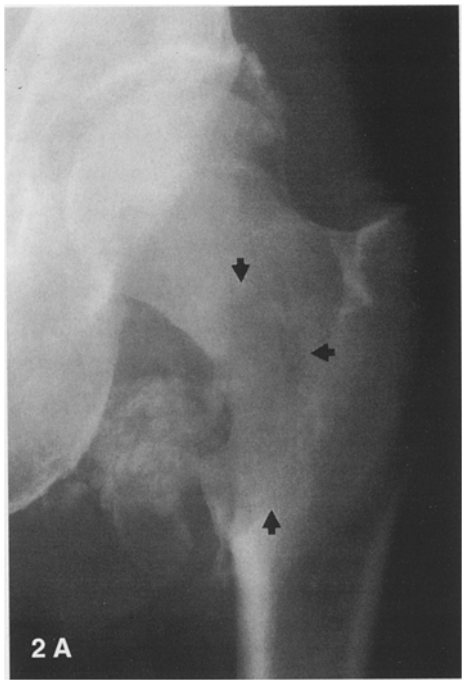
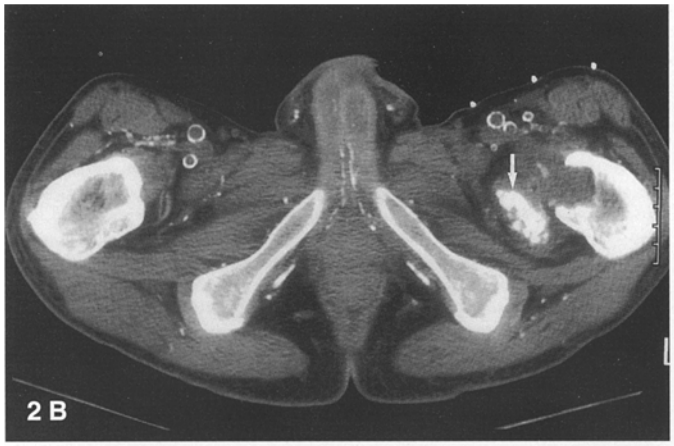
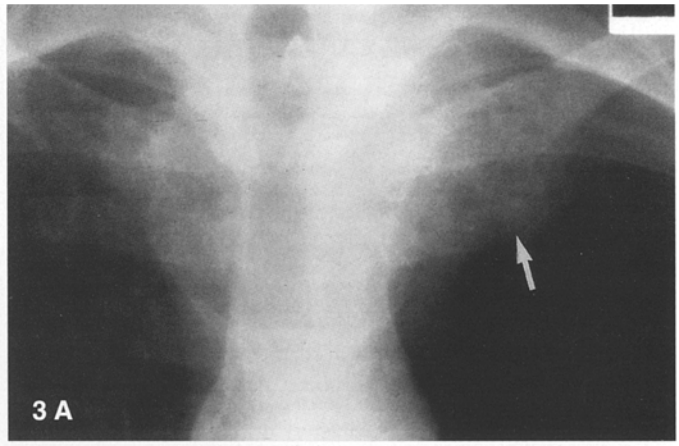


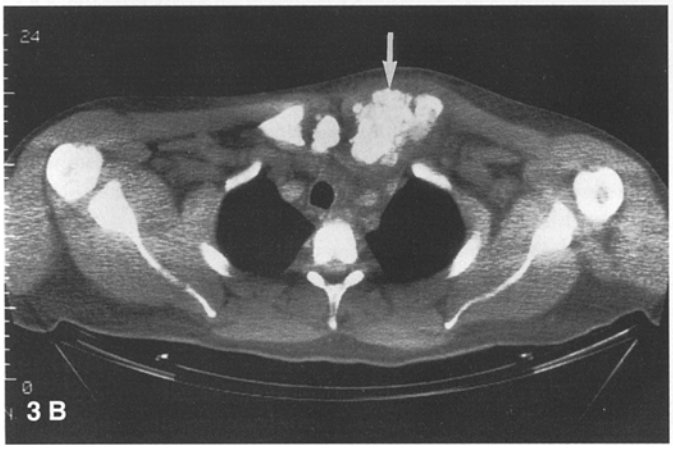
Fig. 2A, B. Case 12. A 58-year-old man was on dialysis and developed pain and a lump in the region of the left hip. He had known tumoral calcinosis of the shoulders and hip which had been previously resected. **A** An AP radiograph of the left hip showed a calcific mass and a large defect in the adjacent bone along the medial femoral neck and lesser trochanter *(arrows),* which was actually a pressure erosion produced by the larger calcified mass that had been removed prior to this study. Note the nodular character of the calcified mass, better appreciated on CT in **B** *(arrow).* The large defect in the anterior femur was found to contain fibrous tissue on biopsy. The lesion was pathologically verified as tumoral calcinosis

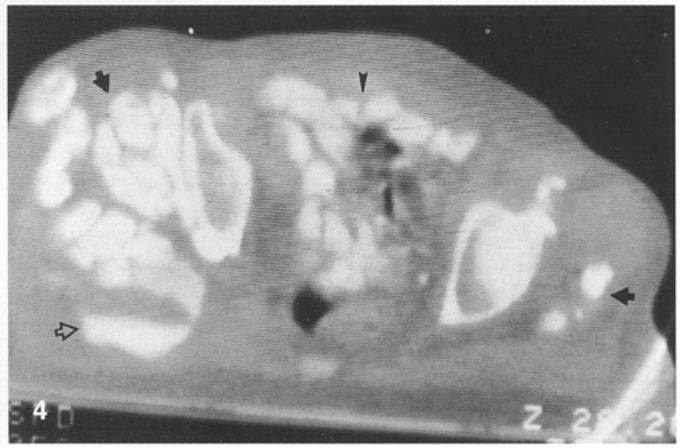
Fig. 3A, B. Case 8. A 30-year-old man with end-stage renal disease had an enlarging mass over the left clavicle seen on **A** a conventional posteroanterior radiograph and **B** CT *(arrow).* It was thought to represent an osteosarcoma or chondrosarcoma preoperatively, and a wide excision was performed with removal of the left clavicle, the right clavicular head, the manubrium, and portions of the first and second ribs

Fig. 4. Case 2. CT scan of a 62-year-old man with primary tumoral calcinosis of the elbows and hips. He was first thought to have a hernia by radiologists who viewed this scan. Note the high-density septated masses of tumoral calcinosis adjacent to the hips *(arrows),* which have a striking resemblance to bowel filled with contrast *(arrowhead).* There is a characteristic calcium-fluid level on the right *(open arrow)*

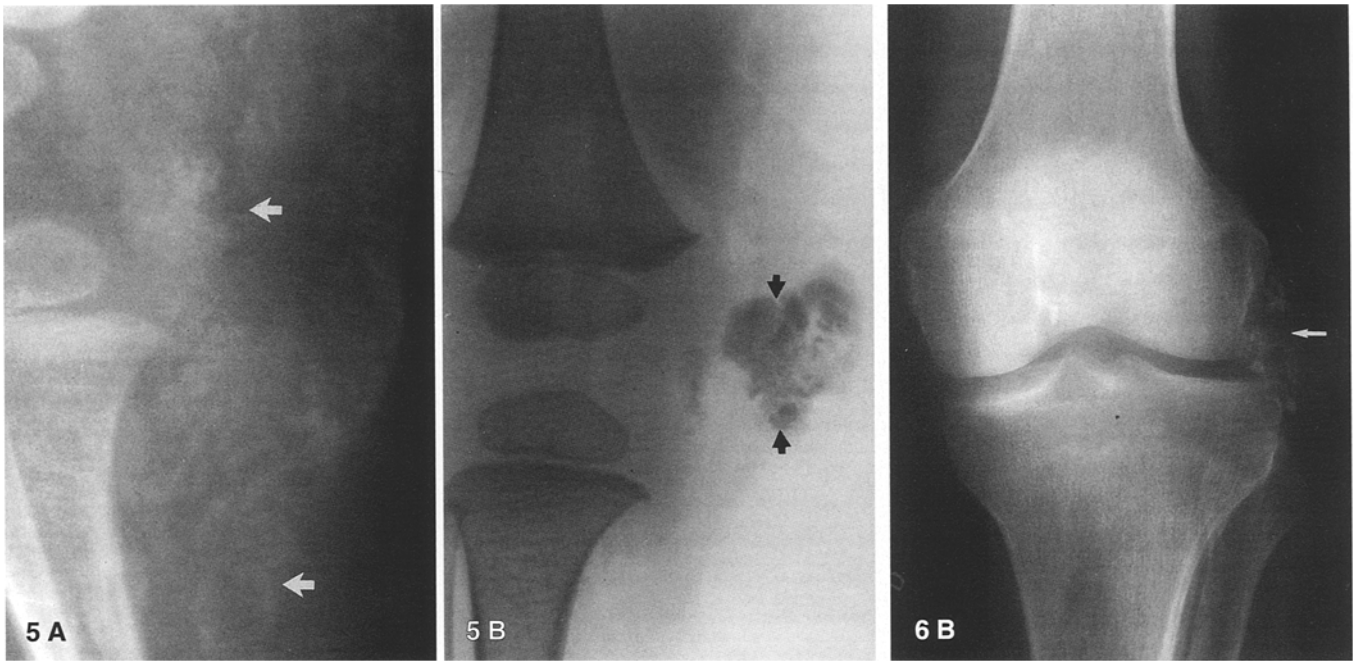








1B). The lobules were filled with amorphous calcareous and occasionally liquefied material. The calcium and fibrous tissue caused the lesions to have a predominantly low signal intensity on T2-weighted MR scans. Fluid-calcium levels were demonstrated in one patient by conventional radiography and in another by CT. Osseous erosion adjacent to the mass was seen in 3 of our patients. One patient had erosion along the undersurface of the humeral head, and another had a large smooth erosion of the femoral neck appearing like a lytic lesion en face (Fig. 2). Osseous involvement was not seen in any of the other cases. Three patients were originally thought to have either osteosarcoma or chondrosarcoma by radiologists and clinicians involved in the cases (Fig. 3).



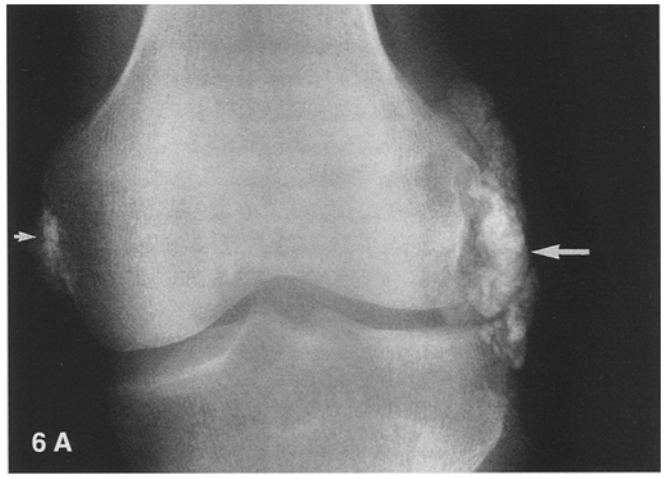


Fig. 5A, B. Case 6. A 15-month-old male with primary tumoral calcinosis of both legs. **A** An AP radiograph of the right leg shows amorphous calcification medial to the knee *(arrows)*. **B** One year later, and without any specific treatment, the lesion coalesced into the more typical nodular pattern of tumoral calcinosis *(arrows)*

Fig. 6A, B. Case 7. A 39-year-old man (other knee shown in Fig. 1) was treated with phosphate binders after he developed secondary tumoral calcinosis around the knees, axilla, and shoulder. **A** Prior to medical therapy there is calcification lateral *(large arrow)* and medial *(small arrow)* to the knee. **B** The calcified masses and are reduced in size and number laterally *(arrow)* and disappear medially following treatment

In one case of idiopathic tumoral calcinosis, the lesion was confused with bowel and thought to represent a hernia on a CT obtained at an outside imaging center (Fig. 4). Another patient was preoperatively diagnosed as having an abscess.

All of the patients except one had surgical resection of the mass. In the one who did not have resection, who happened to be the youngest patient in our series (15 months), the lesion coalesced without treatment, as seen on a 1-year follow-up roentgenogram (Fig. 5). One patient who had resection of a large mass near his right knee had radiographic demonstration of nonsurgical decrease in the calcific deposit following administration of phosphate binders (Fig. 6). Three patients were reoperated on following recurrence of the mass after the first surgical resection.

Seven of the patients had follow-up careful pathologic examination of their specimens by our pathologist (G. H.). A representative example is shown in Fig. 7. The tumor size of the masses was quite variable, ranging from 4 to 23 cm. The fibrous septa were a combination of thick and thin in the same specimen in all patients except the youngest, in whom the septa were exclusively thick. The lobules ranged in size from 0.5 to 14 mm. The lobular lining was a mixture of fibrous and inflammatory tissue in all patients except one, where the lining consisted of purely fibrous tissue.

The lobular contents were analyzed for the presence of amorphous material, mineralized contents, calcospherites, giant cells, and macrophages. All patients demonstrated amorphous material. Mineralized material was present in five. Calcospherites (which arise when the calcified material forms small psammoma body-like masses with concentric layering of calcium which bear a resemblance to ova of parasites) were seen in four patients. Multinucleated giant cells and macrophages were each present in four patients. The mass extended into surrounding soft tissue with fibrosis and inflammatory changes in three patients.

The five comparison cases were also reviewed radiographically and pathologically. Except for the case of scleroderma, none of the comparison cases showed the

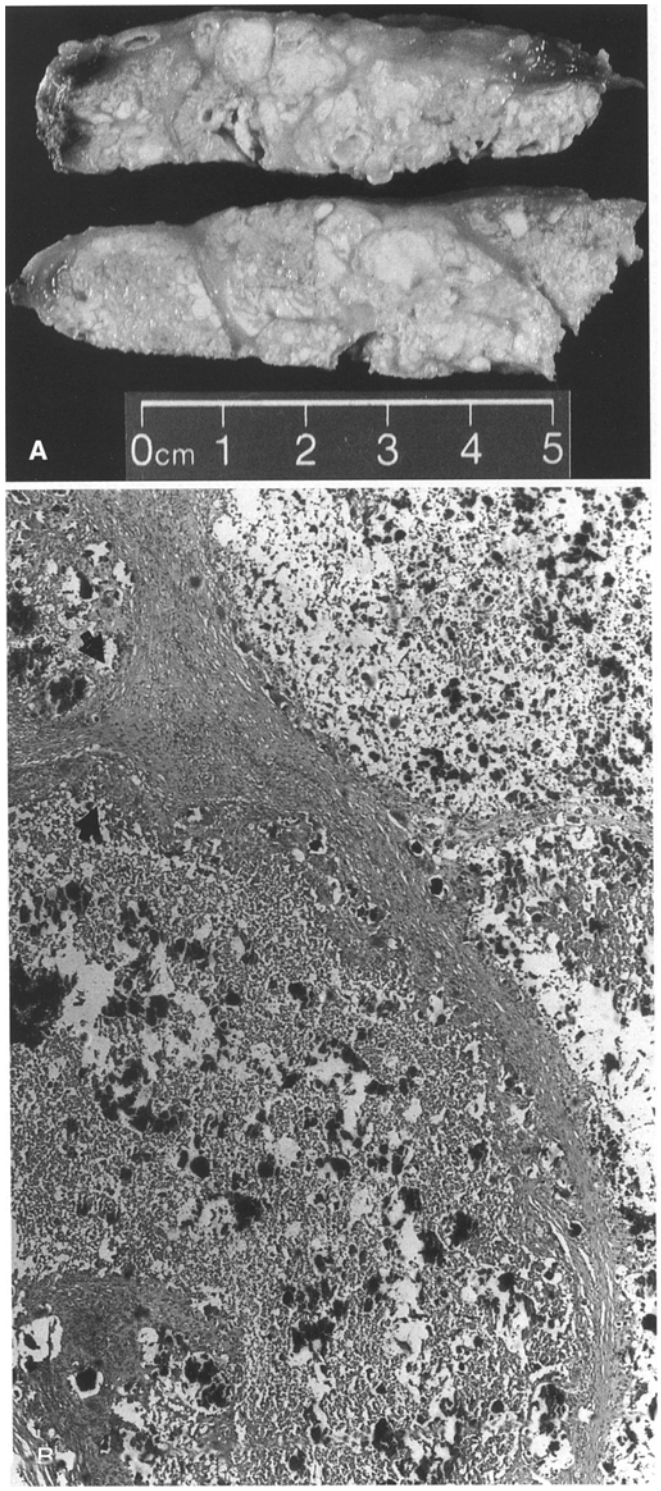


Fig. 7A, B. Case 9. Secondary tumoral calcinosis in a 48-year-old man on chronic renal dialysis. **A** The cut surface of the tumor was yellow-white with a multilobular honeycomb pattern. **B** There are fibrous bands crossing to intersect in one section of the lobule on this histologic section *(arrows)*. Amorphous calcareous debris is seen within the lobule

fibrous septa or lobular configuration seen with tumoral calcinosis. Calcium-fluid levels were not seen in any of the comparison cases (Table 2).

We have observed several cases of scleroderma that have a radiographically similar appearance to tumoral calcinosis, including the case used for comparison in this paper. In that patient, the calcified masses were seen in the hands. There were multiple small homogeneously calcified masses with a "chicken wire" pattern of lucencies that gave them a cobblestone appearance, simulating the radiographic appearance of tumoral calcinosis. Although there was no direct pathologic evaluation of this material, other cases similar to ours have shown that the septa represent thick collagen bundles, and it has been suggested that these septa do look like tumoral calcinosis on gross pathologic examination. However, at the microscopic level, the collagenous septa of scleroderma are thicker than those of tumoral calcinosis (approximately 40 nm), with fewer fibroblasts (personal communication, Phillip LeBoit, M.D.). In our case, the patient's history aided the differentiation of scleroderma from tumoral calcinosis.

Discussion

Tumoral calcinosis is a disease characterized by large calcific periarticular soft tissue masses composed of hydroxyapatite crystals located at multiple sites, usually around large joints such as the shoulders and hips, as in our study, and along extensor surfaces [5]. When it is idiopathic it is seen in younger individuals [6, 7], with a predilection for black people and people from tropical climates [8, 9]. The idiopathic form can also be familial [5, 10–12]. The secondary form of tumoral calcinosis is caused by metastatic calcification. This occurs when there is a high calcium-phosphorus product, a condition which is seen in many patients with renal disease.

The diagnosis of tumoral calcinosis can be missed due to lack of familiarity with its appearance on various imaging studies. Characteristic imaging features were seen in the tumoral calcinosis patients in our study. Some of these features have been addressed in the literature [13–16]. Other features have not been sufficiently emphasized.

All masses of tumoral calcinosis in this study were well defined and had an amorphous lobulated appearance separated by fibrous septa that gave a "chicken wire" appearance to the mass, particularly evident on conventional radiographs and CT. Fluid-calcium levels (also known as the "sedimentation sign") were seen on radiographs and CT in three patients. These comprise semifluid calcified material, similar to milk of calcium bile, with mineral pooling in dependent portions. The supernatant fluid floats above the calcium. Osseous erosion adjacent to the mass was seen in three of our patients. This finding has been rarely described in association with tumoral calcinosis [17, 18].

It is of interest that in five of the patients with tumoral calcinosis, the initial clinical diagnoses made by clinicians and general diagnostic radiologists were the following: osteosarcoma and chondrosarcoma in three patients; herniated bowel on CT in a patient with primary tumoral calcinosis; and an abscess that was clinically diagnosed in another patient with no change in diagnosis following imaging studies. In all of those cases, the imaging findings were characteristic of tumoral calcinosis in retrospect.

There is a large differential diagnosis for tumoral calcinosis, including those processes that produce soft tissue calcification and ossification. These include neoplastic soft tissue calcification or ossification as seen in osteosarcoma, chondrosarcoma, and synovial sarcoma, as well as dystrophic and metastatic calcification or ossification seen in benign soft tissue masses, such as myositis ossificans, heterotopic bone, collagen vascular disease, debris in and around neuropathic joints, tumoral calcium pyrophosphate deposition disease, hydroxyapatite deposition disease, hypoparathyroidism, hypervitaminosis D, and gout.

In the representative cases of other disease processes that can produce soft tissue calcification or ossification presented in this paper, with the exception of scleroderma, there was a distinct difference in the appearance of the soft tissue calcifications, which allowed us to distinguish them from tumoral calcinosis (Table 2). In the case of scleroderma, the calcifications were irregular and inhomogeneous without septation. No calcium-fluid levels were demonstrated. Myositis ossificans demonstrated the zonal pattern of ossification peripherally, which is characteristic of the later stages. Osseous erosion can be a component of other entities that cause soft tissue calcification and cannot be used to distinguish them from tumoral calcinosis.

In scleroderma and dermatomyositis, soft tissue calcifications may be diffuse or focal, and we have observed changes on radiographs similar to those of tumoral calcinosis. The homogeneously calcified masses of scleroderma can also display a cobblestone appearance, similar to tumoral calcinosis, and as in tumoral calcinosis, calcified masses associated with scleroderma can be accompanied by adjacent osseous erosions. The hands are frequently involved in scleroderma, but are not commonly affected in patients with tumoral calcinosis. In these patients, history is helpful for diagnosis, as is the demonstration of characteristic soft tissue resorption of the fingertips and acroosteolysis. However, calcifications of scleroderma can also present in the face, axilla, forearms, lower legs, and ischial tuberosity and can be difficult to distinguish from tumoral calcinosis.

In summary, there are characteristic radiologic findings of tumoral calcinosis that we would like to emphasize. These include the cobblestone or "chicken wire" appearance produced by the amorphous calcium deposits with intervening fibrous septae, seen in the mature tumor. Some patients demonstrate the sedimentation sign, which is also characteristic. Tumoral calcinosis can be associated with bone erosion that can lead to confusion with other tumors. The characteristic appearance of tumoral calcinosis was not demonstrated in four types of calcified masses we examined. We want to emphasize that we have not examined a broad spectrum of calcified and ossified neoplasms. Instead we have tried to illustrate how different these lesions are both radiographically and pathologically from tumoral calcinosis. It is conceivable that occasionally other masses can have a lobular appearance that can be difficult to distinguish from tumoral calcinosis, but in our experience this is not common. On the other hand, we have observed several cases of scleroderma that have a radiographical appearance that is similar to tumoral calcinosis.

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