

## Tumoral Calcinosis Causing Bilateral Thigh Pain

J. Mark Evans, M.D., Catherine C. Roberts, M.D., and Thomas K. Lidner, M.D.

We report a case of a 75-year-old female with bilateral thigh pain for several years secondary to soft tissue calcification. Massive calcinosis of the soft tissues is a unique, but not uncommon, radiographic finding. On the contrary, tumoral calcinosis is a rare familial disease. The term tumoral calcinosis has been overly used to describe any massive collection of periarticular calcification. The original definition of tumoral calcinosis refers to a hereditary disease associated with massive periarticular calcification without an underlying cause. The lesions of tumoral calcinosis are typically lobulated, well-demarcated calcifications most often distributed along the extensor surfaces of large joints. Many conditions have similar radiographic appearances, including the calcinosis of chronic renal failure, calcific tendinitis, synovial osteochondromatosis, synovial sarcoma, myositis ossificans, tophaceous gout, and calcific myonecrosis. The radiologist plays a critical role in guiding the appropriate tests that can result in a conclusive diagnosis of tumoral calcinosis.

### Introduction

The diagnosis of tumoral calcinosis was first described in the American literature in 1943 by Inclan et al (1). Inclan et al differentiated tumoral calcinosis from the dystrophic and metabolic ("metastatic") calcifica-

tions associated with renal osteodystrophy, connective tissue disease, and hormonal imbalance.

In the mid-1960s, reviews established that tumoral calcinosis had a familial tendency without sex predominance but with a significantly higher incidence in patients of African descent (2). Lesions primarily proliferate during the first few decades of life. Although all of the studied patients had normal serum calcium levels, a minority of patients had mild hyperphosphatemia. The classic tumoral calcinosis lesions were described as lobular, densely calcified masses confined to the soft tissue, generally at the extensor surface of the joint. In 1990, Martinez et al (3) described additional characteristics using bone scintigraphy, computed tomography (CT), and magnetic resonance (MR) imaging. The most common locations of tumoral calcinosis in descending order are the hip, elbow, shoulder, foot, and wrist.

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

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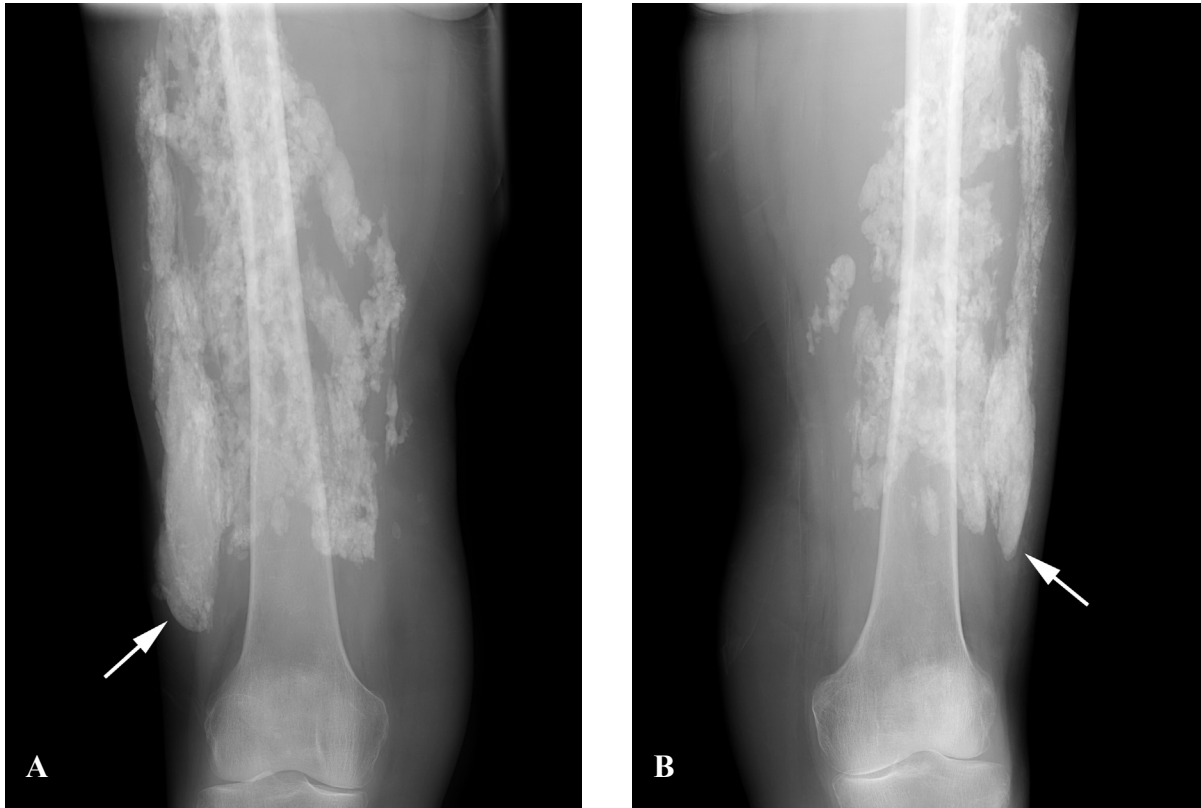


Figure 1. 75-year-old woman with bilateral thigh pain. Radiographs of the right (A) and left (B) femora demonstrate extensive regions of soft tissue calcification (arrows) in the thighs.

### Case Report

A seventy-five-year-old female presents with a five year history of increasing fullness and pain within both thighs. She had undergone surgical treatment in the past with subsequent wound complications and infection. Her past medical history is significant for sarcoidosis. The patient had no evidence of a mixed connective tissue disorder or renal disease.

On physical examination, large firm masses were palpated in the anterolateral thighs bilaterally. The masses were nontender. Her neurovascular examination of the lower extremities was normal. Her hip and knee examination revealed a full range of motion. Her calcium and phosphorus levels were within normal limits.

Radiographs (Figure 1) of both femora demonstrate masses of soft tissue calcification in the thighs without cortical erosion or mass effect. Radiographs of both

humeri (Figure 2) also demonstrate large areas of calcification in the soft tissues without abnormality of the adjacent humeri. Computed tomography of the bilateral thighs (Figure 3) shows the calcifications are symmetric and most extensive in the distal thirds of the thighs.

The patient was initially treated conservatively with restrictions in her dietary intake of phosphorus. Surgical intervention was avoided due to her history of postoperative infection.

After two years of conservative treatment, she presents with increasing pain and erythema over the lateral aspect of her distal right thigh. Magnetic resonance imaging (Figure 4) of her right knee demonstrates a mass-like area of calcification in the vastus lateralis muscle, unchanged from the calcification seen on the previous radiographs. The patient agreed to continue with conservative therapy.

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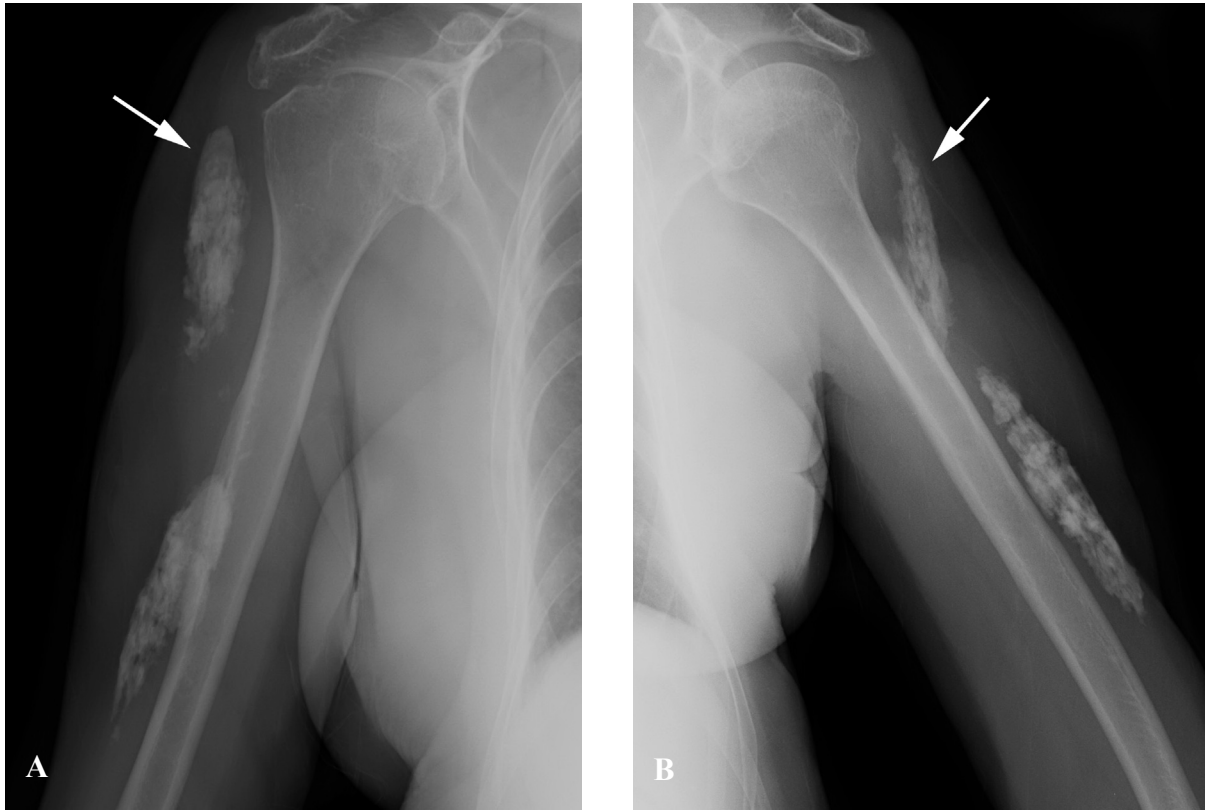


Figure 2. Radiographs of the right (A) and left (B) humeri demonstrate extensive calcified soft tissue deposits (arrows).

She presents one year later with increasing pain in her anterolateral distal right thigh. Her physical examination reveals severe edema and erythema at the site of pain. Computed tomography of the right thigh (Figure 5) reveals the mass-like area of calcification and soft tissue density in the distal right vastus lateralis muscle has increased in size and is significantly closer to the skin surface. This area was surgically excised due to the risk of impending rupture.

The surgical pathology revealed fibrous tissue and calcified debris (Figure 6). The gross examination revealed a cystic structure filled with gray-tan, partially calcified, putty-like material measuring 7 x 5 x 4 cm. The histological features were not specific, but consistent with the clinical diagnosis of tumoral calcinosis.

Her pain and swelling improved significantly post-operatively. There were no complications. IRB approval for case reports is waived at our institution.

### Discussion

Tumoral calcinosis refers to a non-aggressive appearing calcific mass often centered around large synovial joints. This is a rare entity and is commonly misdiagnosed. The most commonly involved joints are the hip, shoulder, and elbow. Spinal involvement can occur but is relatively uncommon. The size of tumoral calcinosis is quite variable, but inevitably presents as a well defined mass or cluster of masses.

### *Pathogenesis*

The exact etiology of tumoral calcinosis remains unclear with several proposed theories. One of these theories is that patients may have inborn errors of phosphorus metabolism, which predispose them to tumoral calcinosis (4). A second theory is that hemorrhage from micro trauma causes an exaggerated reparative response.

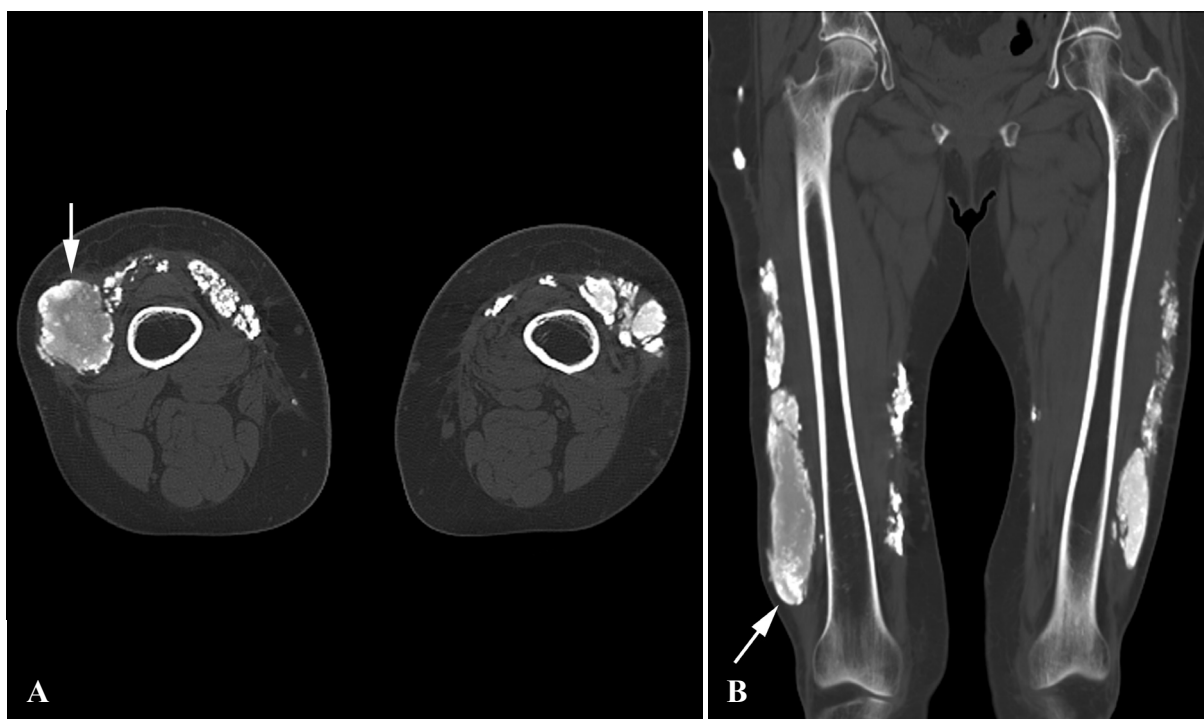


Figure 3. Axial (A) and coronal (B) computed tomography of bilateral thighs shows that the calcified masses (arrows) within the quadriceps muscles are relatively symmetric and most extensive involving the distal thirds of the thighs.

A third theory is that tumoral calcinosis represents a variant of calcium pyrophosphate deposition disease.

In the literature, tumoral calcinosis has been categorized as either primary or secondary. Primary tumoral calcinosis referred strictly to a disease caused by a hereditary metabolic dysfunction of phosphate regulation. In contrast, secondary tumoral calcinosis referred to calcified masses associated with an identifiable condition, most often chronic renal failure.

The term tumoral calcinosis has been liberally and imprecisely used to describe any massive collection of periarticular calcification. The inconsistent use of this term has created confusion throughout the literature.

The classic definition of tumoral calcinosis refers strictly to a disease caused by a hereditary metabolic dysfunction of phosphate regulation associated with massive periarticular calcinosis (1).

One approach to differentiating tumoral calcinosis from its mimics is by categorizing soft tissue calcification in terms of serum chemistry levels. Metabolic calcification usually results in generalized mineral deposition, in-

cluding visceral organs, and is associated with abnormal calcium and/or phosphate levels. Dystrophic calcification results from an underlying inflammatory process and is found in patients with normal serum chemistry levels. With normal serum levels, antibody screening for an underlying rheumatic disease is recommended. Finally, idiopathic calcification, which refers to tumoral calcinosis, is associated with normal calcium and elevated/normal phosphate concentrations.

The etiology of our patient's soft tissue calcification remained indeterminate, and the diagnosis of "tumoral calcinosis" was established. An atypical feature in our patient is that the soft tissue calcification is more muscular than periarticular.

## Differential diagnosis

### Dystrophic calcification (4-5)

1. Connective tissue diseases such as scleroderma and dermatomyositis
2. Neoplasms such as synovial sarcoma and chondrosarcoma

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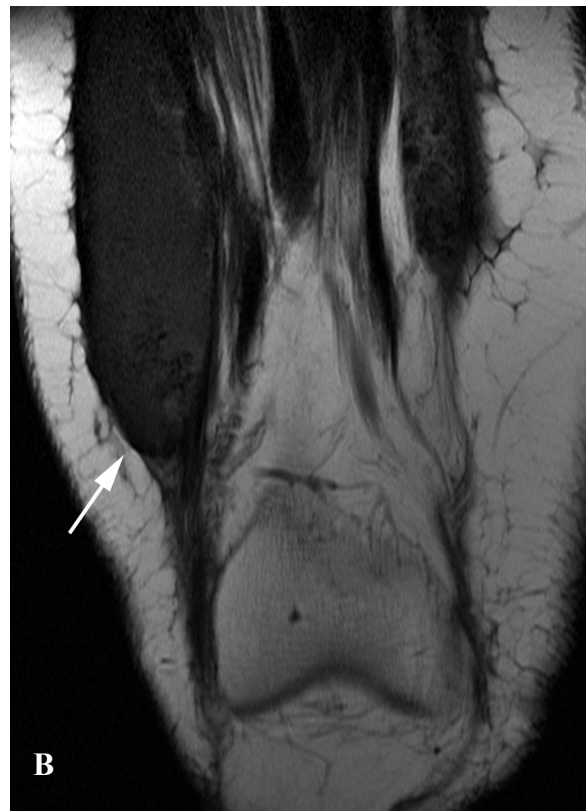


Figure 4. Axial T1-weighted (A), coronal T1-weighted (B) and coronal inversion recovery (C) magnetic resonance images of the right knee show the mass-like area of calcification (arrows) in the vastus lateralis muscle. The mass has heterogeneous isointense to hypointense signal on T1 and homogeneous low signal on the inversion recovery sequence.



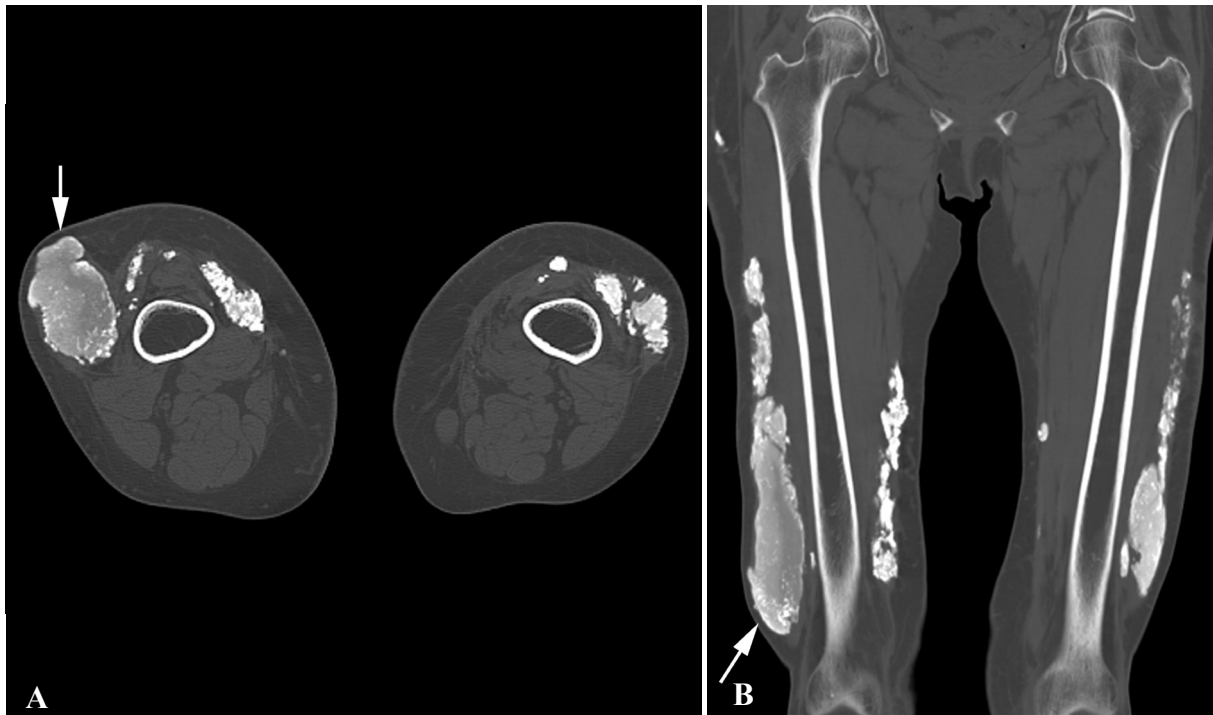


Figure 5. Axial (A) and coronal (B) computed tomography of the bilateral thighs reveals the mass-like area of calcification (arrows) in the distal right vastus lateralis muscle had increased in size over the intervening two years (See Figure 3 for comparison).

3. Synovial osteochondromatosis
4. Calcium pyrophosphate deposition disease, calcific tendinitis, and calcific bursitis

#### Metabolic calcification (4-5)

1. Hyperphosphatemia secondary to chronic renal failure
2. Hypercalcemia from primary hyperparathyroidism and milk alkali syndrome
3. Hyperuricemia secondary to tophaceous gout

#### Imaging

With radiography, tumoral calcinosis has the typical appearance of amorphous, cystic, and multilobulated calcification located in a periarticular distribution (6). There may occasionally be bone remodeling without destruction. These findings are also well demonstrated on computed tomography. The computed tomography (CT) also shows the absence of a noncalcified soft tissue mass. With magnetic resonance imaging (MRI), tumoral calcinosis most often shows lobulated low signal

masses on both T1-weighted and T2-weighted sequences. Following gadolinium, tumoral calcinosis demonstrates minimal enhancement without an enhancing soft tissue component. With bone scintigraphy, increased radiotracer uptake of these lesions is most often demonstrated (3).

The best imaging tool to evaluate calcified masses is computed tomography (CT) as this modality most accurately shows calcified lesions and the presence or absence of bone destruction.

#### Pathology

The pathologic and surgical features are a calcific mass with a mixture of solid and “milk of calcium” components. The gross pathologic features typically show an unencapsulated mass with extension into tendon and muscle. Sectioning shows dense fibrous tissue surrounding spaces filled with pasty calcified material. Microscopically, amorphous calcified material is surrounded

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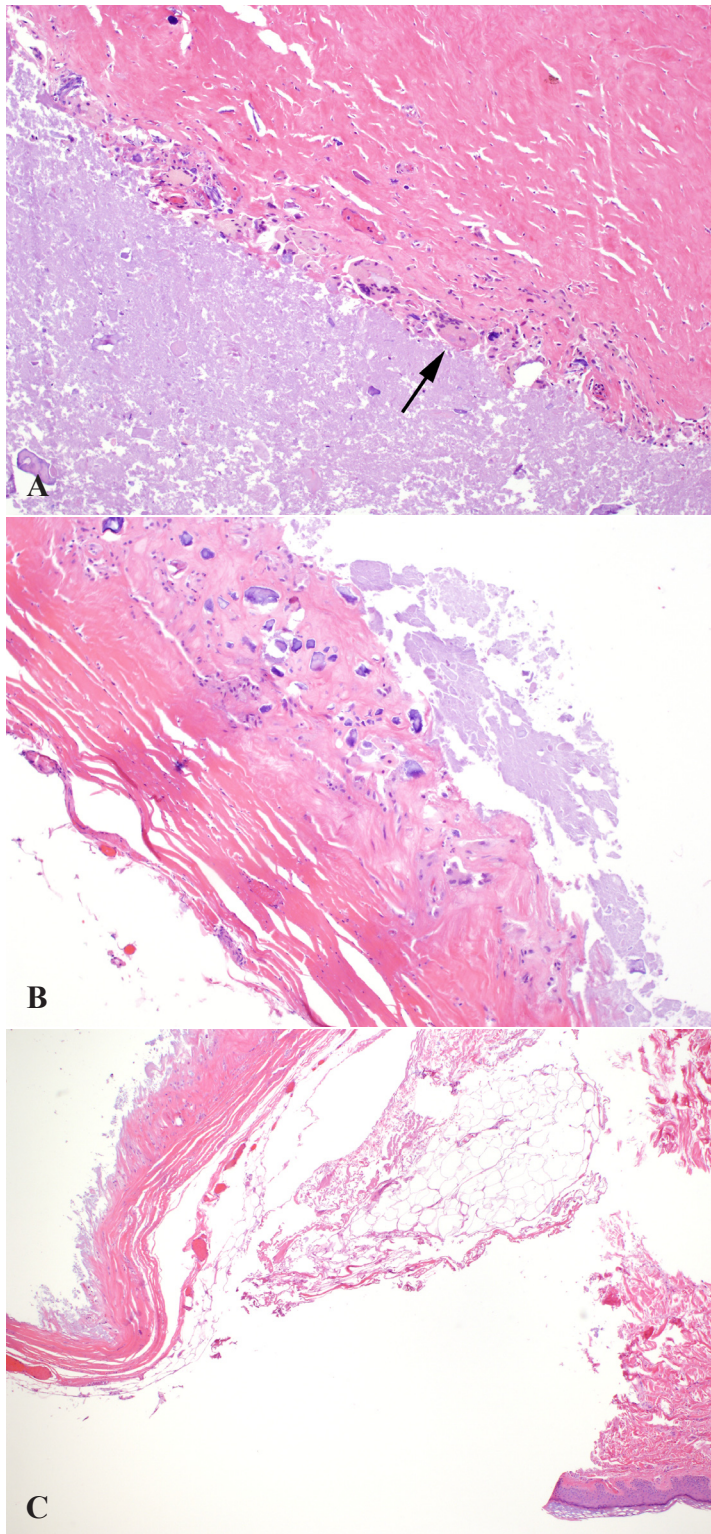


Figure 6. Histologic section (Hematoxylin-eosin; x100, x100, x40 respectively) photographs of the surgically excised mass shows (A) amorphous calcified material bordered by a few osteoclast-like giant cells (arrow) with a surrounding wall of fibrous tissue, (B) amorphous calcified material with surrounding fibrous tissue, and (C) the cystic fibrous-walled cavity beneath the skin.

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by dense fibrous tissue. Calcium hydroxyapatite and calcium pyrophosphate dihydrate crystals are often identified. The activity of the disease varies, sometimes within the same lesion. More active cases demonstrate more numerous macrophages, fibroblasts and osteoclast-like giant cells (7).

### Treatment

The treatment of massive soft tissue calcification depends largely on its underlying cause. Surgical excision of the lesions of tumoral calcinosis is a well documented treatment; however, recurrences due to incomplete resection are common. Surgical excision combined with phosphate deprivation (using aluminum hydroxide) and acetazolamide therapy synergistically lowers hyperphosphatemia and has proven to be the most effective therapy (8). Other therapies such as systemic steroid therapy and radiation therapy have not proven effective.

Metabolic calcification is best treated by addressing the underlying cause of the metabolic dysfunction. For example, hemodialysis is used for patients with renal osteodystrophy and hormonal regulation is the treatment for primary hyperparathyroidism.

### Conclusion

Tumoral calcinosis is a hereditary disease of phosphate metabolic dysfunction. This dysregulation leads to the formation of characteristic, lobulated, well-demarcated calcification distributed most commonly around the extensor surface of large joints. Because there are many conditions with similar-appearing lesions, formulating the exact diagnosis is sometimes difficult with imaging alone. However, the radiologist plays a critical role in decision making to avoid unnecessary procedures and provides the consulting physician with direction in selecting the appropriate tests for a conclusive diagnosis.

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