

Tumoral Calcinosis With Renal Failure: Case Report

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Introduction

Massive deposition of calcium salts presenting as tumors are rarely encountered. However, they are known to occur in patients with kidney failure, hyperphosphatemia, and secondary hyperparathyroidism. It is not clear why most patients with this condition only develop discrete calcifications of the smaller vessels, the cornea, mitral valves, and other disseminated areas. In addition to disseminated calcinosis, some patients will develop focal calcium tumors that, by virtue of their size, cause severe pain, soft tissue destruction, and occasionally local infections.

We present a patient with large calcific masses, for whom it was necessary to include an orthopaedic surgeon on the team caring for her.

Case Report

A 71-year-old white female was admitted to the hospital in January 1997 with a one-week history of increasingly severe pain in the right thigh and slightly less pain in the left thigh. The patient had a long history of coronary artery disease and suffered a myocardial infarction and repair of an abdominal aneurysm in 1991. At the time of this admission, she was under treatment for renal failure, hypertension, and peripheral vascular disease, which had required an above-knee amputation on the right in 1993. Her reduced kidney function was due to atheroembolic disease and required dialysis three times a week since 1993. In 1994, a healing fracture of the left clavicle was noted on a routine chest film and was associated with a calcific mass in the surrounding soft tissues.

She was recently discharged from another hospital where she had been treated for ten weeks with intravenous antibiotics for a bacteremia resulting from an infection from her permacath. The patient is the mother of three children and has no family history of renal disease or abnormalities of calcium metabolism; she gave up smoking six years ago. Since her amputation, she has remained wheel chair dependent.

Physical examination revealed the patient to be in acute distress, complaining of severe pain in the upper lateral right

thigh with lesser pain in the left thigh. An irregular, circumscribed, firm mass was palpable in the right lateral thigh, about 15 cm in diameter, over which the skin was edematous with intradermal blisters. The swollen area was warm and exquisitely tender. Movements of the right hip joint were not painful and palpation of the femur from the medial side and the femoral stump did not elicit any bone tenderness.

On the left side, a smaller swelling was palpable in the midlateral thigh, which was less tender and the skin was less edematous. Movements of the left hip, knee, ankle, and foot did not cause pain, nor was there any tenderness on palpation of the femur.

The patient's temperature on admission was 38°C (99.7°F), blood pressure 102/60 mmHg, pulse 110 and irregular; the lung fields were clear. The pertinent blood studies were as follows: hemoglobin 8.6 g/dl, white blood count 13.5 K/cm with 90% segmented forms, blood urea nitrogen 39 mg/dl (normal 6–26), creatinine 4.3 mg/dl (normal 0.4–1.5), phosphorus 7.7 mg/dl (normal 2.2–4.3), calcium 7.7 mg/dl (normal 8.7–10.5), pH 7.18 (normal 7.35–7.45). Blood cultures were negative on several occasions, but the patient was receiving vancomycin and gentamicin. Several parathyroid hormone levels on an admission two weeks previously were between 320 and 139 pg/ml (normal 12.1–60.6).

Roentgenogram of the ribs revealed a calcific mass (Fig. 1). Films of the right femur showed severe osteopenia, with a normal appearing hip joint and an undisplaced fracture in the subtrochanteric area of the femur. A lobulated, irregular, calcific mass measuring 17 × 12 cm was lateral to the femoral shaft (Fig. 2). The left femur showed two smaller similar masses, one anterior and one posterior to the upper femoral shaft.

The primary problem for which the orthopaedic surgeon was consulted was to relieve the pain. To accomplish this, it was necessary to consider the fracture, infection, or the bulk of the tumor as the source of the severe pain. As movements of the right limb caused no pain, the fracture was not regarded as the main source of pain. A decision was made to remove the mass on the right and the larger mass on the left.

When a lateral incision was made over the right thigh mass, copious chalky fluid exuded. On incision of the ilio-tibial tract, a large calcified mass, within a pseudocapsule, which was not attached to any structure, was removed. The mass, which was removed, had eroded the lateral femoral cortex and thinned the vastus lateralis muscle. Cultures were

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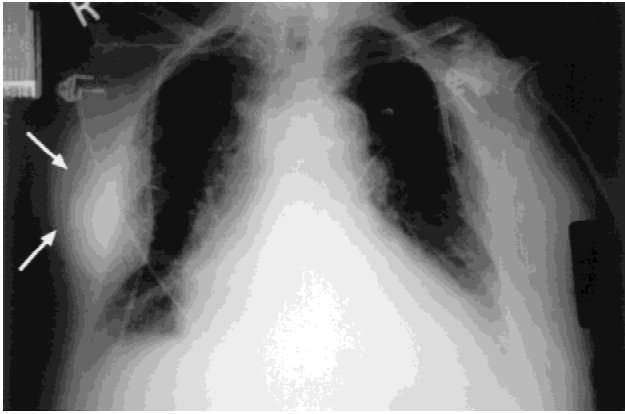


Fig. 1. Calcified mass on ribcage.

taken and additional chalky material was removed with pulse irrigation. The wound was closed in a single layer with drains in situ. The same procedure and similar findings were made in the left thigh.

Postoperatively, the patient was dialyzed and her complaints of pain were minimal. Unfortunately, on the first postoperative day, she developed a seizure, became unresponsive, and died. The clinical cause of death was given as cardiac failure with possible sepsis.

Cultures from both wounds showed no growth. Pathological examination of the larger mass showed it to measure 14.5×6.9 cm. On sectioning, gritty pale yellow tissue was described. Microscopic examination showed amorphous deposits separated into lobules by fibrous septa that were heavily stained with hemosiderin and showed numerous foreign body giant cells (Figs. 3, 4).

A specimen of the tumor sent for crystallographic analy-

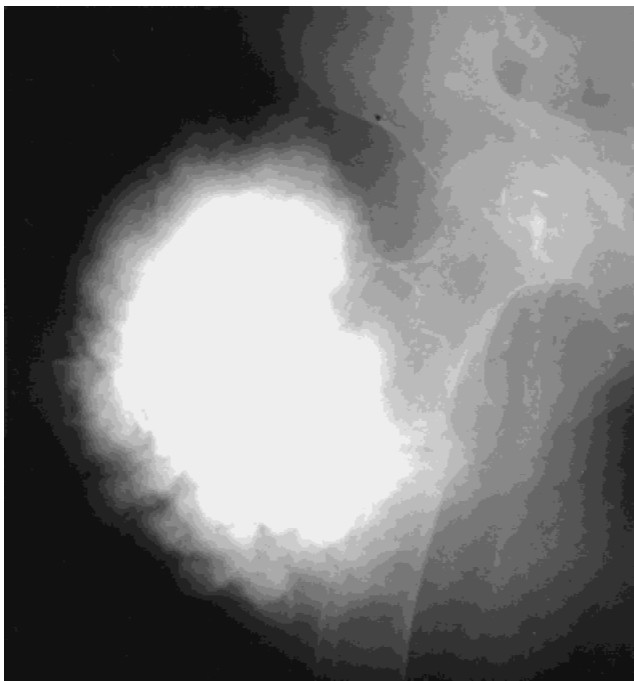


Fig. 2. Right femur showing an irregular calcified mass with a flare, based on an osteoporotic femur. An undisplaced pathological fracture is present.

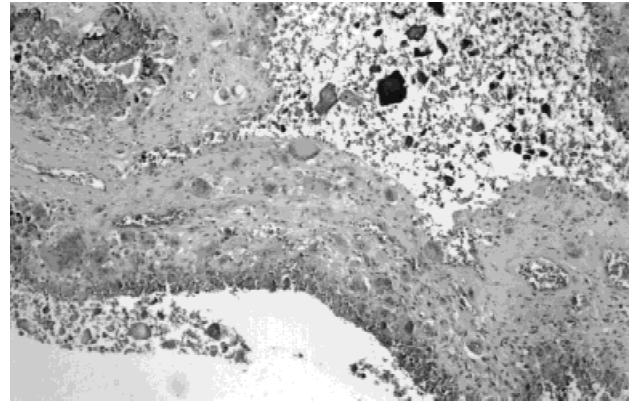


Fig. 3. Microscopic section of specimen (low power) (color) showing fibrous septum separating lobules of calcium (most of the calcium has been leached out by decalcification in preparation).

sis showed numerous irregularly shaped calcific fragments composed of admixed masses of microcrystalline carbonate apatite and hydroxyapatite with proteinogenous tissue and blood and protein matrices. Analysis showed carbonate apatite 80%, hydroxyapatite 10%, and protein, blood, and tissue 10%.

Discussion

Calciophylaxis or disseminated deposition of calcium in tissue is frequently seen in patients with renal disease with or without hemodialysis. When the glomerular filtration rate is below 25% of normal, a rise in serum phosphate and a decrease in serum calcium with secondary hyperparathyroidism and a reduction of the vitamin D metabolite 1, 25 dehydroxyl D_3 occurs. These electrolytes play a major role in bone formation and resorption. With electrolyte abnormalities, the latter process becomes dominant and osteomalacia and osteoporosis result. Other subtle factors, including malfunction of intestinal absorption, dietary alterations, disuse, and aluminum concentration and toxicity, augment the process [1,4,5]. Patients also suffer from small artery disease with calcium deposits in the media of the vessels combined with intimal hyperplasia, which can cause skin necrosis, gangrene, or infection [3]. Uremic patients also de-

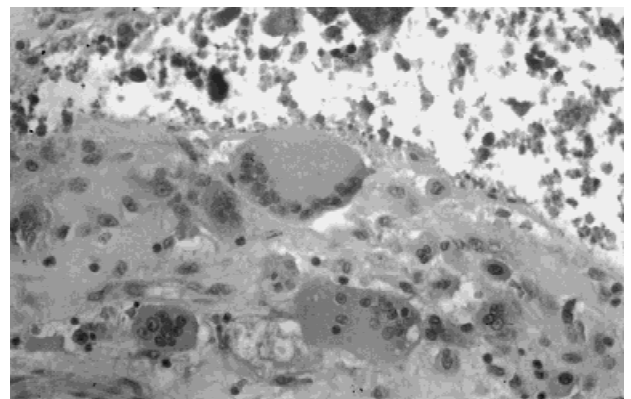


Fig. 4. Microscopic section (high power) of specimen showing a fibrous septum with giant cells and granulation tissue (color).

posit B₂ microglobulin amyloid deposits in many tissues, causing a variety of clinical conditions, including carpal tunnel syndrome.

Tumoral calcinosis is the term used to identify patients with large masses of calcific deposits in the soft tissues. The term is applied to two separate unrelated clinical conditions. In adolescents and young adults, tumor masses of calcium salts are occasionally seen clustered around the hips, shoulders, elbows, and other areas without any disturbance of renal function and usually without changes in calcium metabolism, although hyperphosphatemia is noted occasionally. The etiology is obscure, but it may be related to a genetic predisposition or trauma [2]. When the mass is so large that function is impaired, surgical removal may be necessary.

Tumoral calcinosis associated with renal failure with large deposits of extraosseous masses of calcium crystals is also related to the calcium, phosphorus product, but is rarely encountered. McGregor et al. [6] reported two patients with tumoral calcinosis who had chronic renal failure. In one patient, a calcified multiloculated tumor 20 cm in diameter was located in the gluteal region; in the other, a smaller tumor in the foot recurred after excision. Microscopic examination of their specimens showed compacted hydroxyapatite crystals in a fibrous capsule containing many histiocytes and giant cells. In another report, a patient receiving ambulatory peritoneal dialysis for seven months developed a tumoral calcinosis encasing the extensor tendons of the wrist with loss of wrist function. Surgical removal of the mass restored tendon function [8].

Medical treatment has not been successful in reducing the size of the tumors or controlling the resulting pain. Parathyroidectomy was tried in severe cases by Roe et al. [7]. They reported that only three of their survivors had a minor reduction in tumor size and concluded that prognosis remained dismal for these patients.

Sperschneider et al. [9] treated patients with calcium carbonate for a period up to three years in an attempt to lower

the serum phosphate. Although there was some initial success, it could not be maintained and new masses appeared during the period of observation.

Tumoral calcinosis with renal failure must be differentiated from calcium masses arising from chronic vitamin D intoxication, milk-alkali syndrome, primary hyperparathyroidism, fibro-osseous dysplasia progressiva, calcinosis circumscripta, heterotopic ossification, and myositis ossificans.

In our patient, severe pain that was unresponsive to medication was the primary complaint. The pain was present on both thighs, but was more severe on the right side with the larger mass. We believed that the bulk of the expanding mass with edema of the surrounding tissue was responsible for the pain and this was the basis for the decision for surgery. The presence of calcium deposits as a possible source of pain in patients with renal disease should be familiar to orthopaedic surgeons who may be called upon to assist in the treatment of these patients.

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