

Secondary Tumoral Calcinosis with Intraosseous Penetration

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

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Abstract

We report a case of a 55-year-old woman with chronic renal failure and secondary tumoral calcinosis with rare intraosseous penetration of the humerus. Typical lobulated calcifications with associated layering “milk of calcium” were present about the right shoulder and elbow on conventional radiographs, computed tomography images and magnetic resonance images. Rare appearance of cortical destruction and intraosseous extension of tumoral calcinosis into the medullary space was observed in the proximal humerus on CT and MR images. Since there is concern for pathologic fracture, radiologists should be aware of this rare complication of chronic renal disease.

Case Report

A 55-year-old white woman with long standing history of diabetes and end-stage renal disease presented with a two month history of enlarging right arm mass. She reported having soft tissue masses around right shoulder and elbow for many years but was concerned about recent enlargement of the elbow mass. She denied trauma to this region. Physical examination revealed large lobulated masses that were palpable in the region of the left olecranon bursa, proximal biceps muscle, and proximal triceps muscle. Limited flexion and extension of the elbow was noted but there were no neurological or vascular deficits.

Upright radiographs of the humerus showed large calcified masses adjacent to the right elbow, shoulder, and chest wall. The masses were well-demarcated, lobulated and contained layering “milk of calcium” characteristic of tumoral calcinosis (Figure 1). CT images of the upper extremity confirmed the presence of multiple lobulated masses containing fluid-calcium levels. Particularly interesting was the presence intraosseous penetration of the proximal humerus by the calcified mass. Focal destruction of the anteromedial cortex of the proximal humerus was accompanied by

calcifications extending into the medullary cavity. The intramedullary calcifications contained multiple fluid-calcium levels (Figures 2 and 3). A second collection of soft tissue calcifications was seen within the right chest wall. There was cortical erosion of anterolateral second rib by the calcified mass (Figures 4a and 4b). A third collection of soft tissue calcifications was seen in the posterolateral chest wall at the level of the right 5th and 6th ribs but no osseous penetration was noted. MR images of the humerus showed large lobulated soft tissue masses containing calcifications that were hypointense on T1-weighted and T2-weighted sequences. The characteristic fluid-calcium levels were seen with the fluid layer appearing isointense on T1-weighted and hyperintense on T2-weighted sequences and the “milk of calcium” layer appearing hypointense on T1-weighted and T2-weighted sequences (Figure 5).



Figure 1. 55-year-old woman with secondary tumoral calcinosis. Erect AP radiograph of right humerus shows well-demarcated lobulated masses containing layering milk of calcium about right elbow, shoulder, and chest wall.

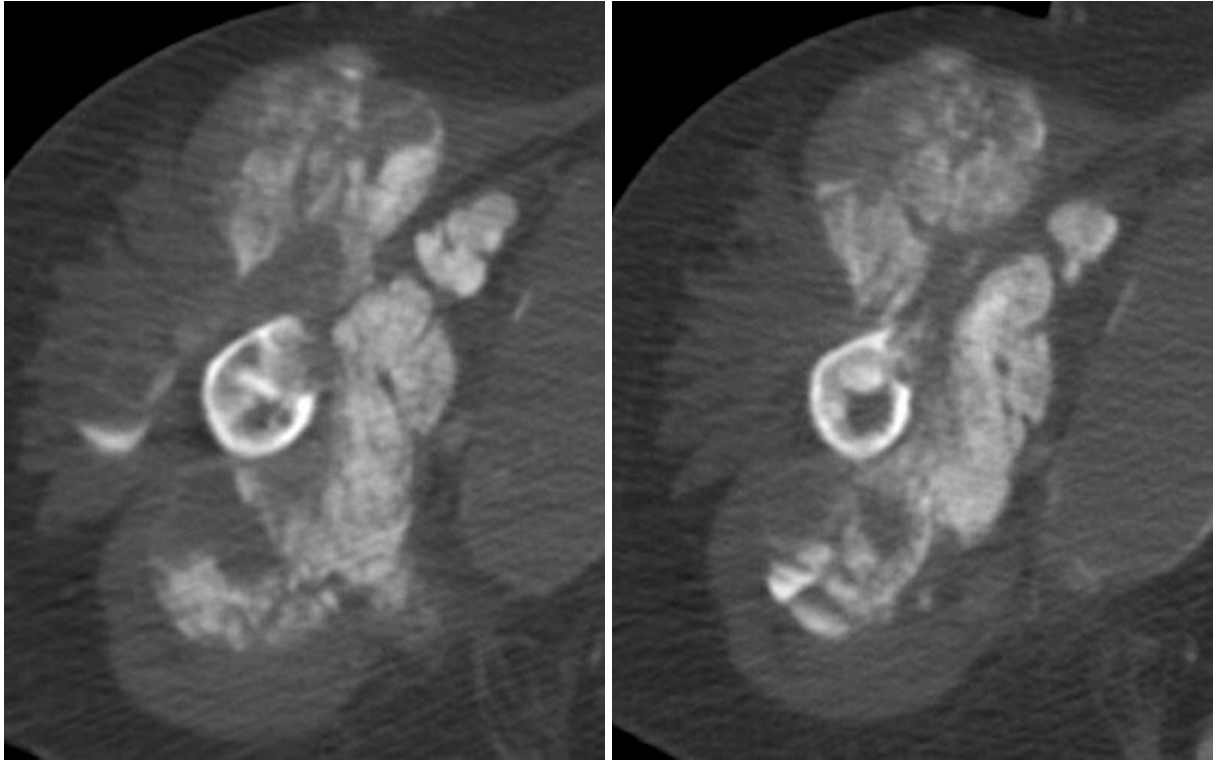


Figure 2. 55-year-old woman with secondary tumoral calcinosis. **(A-B)** Axial CT images of the right upper extremity show lobulated soft tissue masses with multiple fluid-calcium levels. Note cortical destruction of the proximal humerus by tumoral calcinosis with extension into the intramedullary cavity.



Figure 2. (Cont'd) **(C)** Coronal CT image of the right upper extremity shows lobulated soft tissue masses with fluid-calcium levels. Note cortical destruction of the proximal humerus by tumoral calcinosis with extension into the intramedullary cavity.

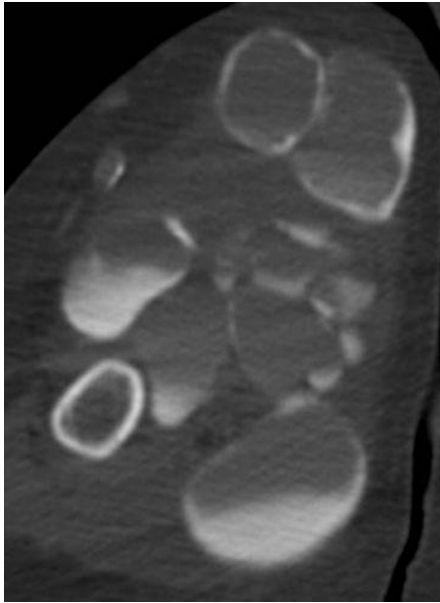


Figure 3. 55-year-old woman with secondary tumoral calcinosis. Axial CT image of the arm shows multiloculated cystic lesions of tumoral calcinosis with fluid-calcium levels.

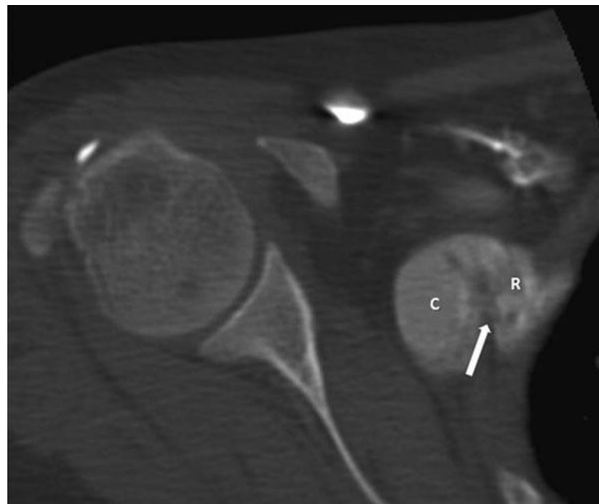


Figure 4A. 55-year-old woman with secondary tumoral calcinosis. (A) Axial CT image of the right upper extremity showing soft tissue calcification (C) with bony erosion (arrow) of the right second rib (R).

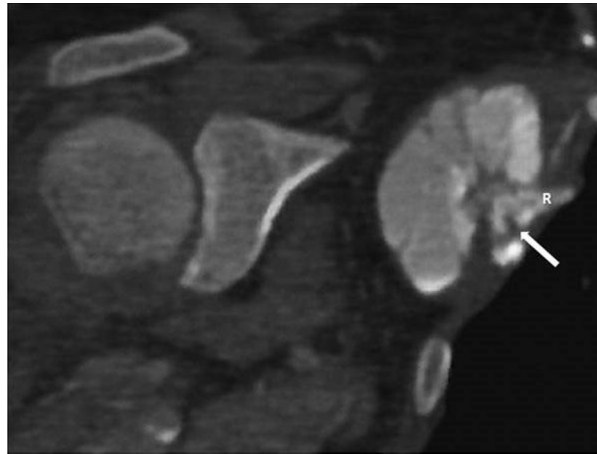


Figure 4B. 55-year-old woman with secondary tumoral calcinosis. **(B)** Coronal CT image of the right upper extremity showing bony erosion (arrow) of the right second rib (R).

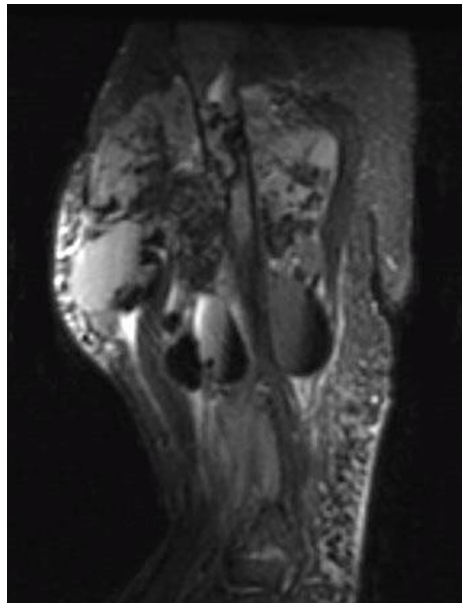


Figure 5. 55-year-old woman with secondary tumoral calcinosis. Sagittal STIR MRI shows intraosseous penetration into the intramedullary cavity of the proximal humerus and fluid-calcium levels.

Discussion

Tumoral calcinosis was first described by Giard [1] and Duret [2] in the 1890s. Tumoral calcinosis refers to lobular, calcified soft tissue masses that occur in periarticular locations, especially the hip, elbow, shoulder, foot and wrist [3]. The masses are often multiple and bilateral [4]. Tumoral calcinosis is usually classified as primary (idiopathic or familial) or secondary [5]. The primary form typically presents with soft tissue masses in the first or second decade of life, has no gender predilection, and is more common in blacks [5-6]. The secondary form is associated mainly with chronic renal failure, has 0.5% - 1.2% prevalence in hemodialysis patients, and has been attributed to secondary hyperparathyroidism [7-8]. In renal patients, the masses typically grow slowly over months or years and may become symptomatic as they produce mass effect on adjacent structures.

On radiographs, tumoral calcinosis appears as multilobulated calcific deposits in a periarticular distribution, usually along the extensor surfaces of joints [6]. The calcified masses may have a dense appearance or may contain fluid-calcium levels, known as the “sedimentation sign” [9]. Bone destruction and erosive changes are characteristically absent, but have been reported [10-12]. On CT images two distinct patterns have been described: 1) cystic collections with low attenuation centers and calcific walls, containing fluid-calcium levels and 2) multilobulated masses with uniform calcifications. The first pattern is considered indicative of a more metabolically active lesion whereas the second pattern is considered more metabolically stable [4]. On MR images, the calcified masses have predominantly low signal on T1-weighted and T2-weighted sequences. Two MR patterns were reported by Martinez et al [4]: 1) a nodular pattern with areas of mixed high signal intensity and signal void and 2) a more diffuse lower signal intensity pattern.

The treatment options for patients with tumoral calcinosis include conservative therapy with phosphate depletion medications. If conservative therapy fails, subtotal parathyroidectomy may be considered [13]. Occasionally, wide surgical excision may be performed, but recurrence is common.

Histopathological examination of tumoral calcinosis shows calcified masses with foreign body inflammatory reaction and fibrotic encapsulation [12, 14, 15]. Septa divide cystic components which contain fluid and granular basophilic material. The walls of the cysts contain dense hyalinized fibrous tissue with some lymphocytes, plasma cells, and macrophages [14].

Bone involvement by tumoral calcinosis is extremely rare but has been reported. Meltzer et al [10] reported a case of a 73-year-old black woman on chronic hemodialysis with tumoral calcinosis eroding the iliac crest. Meneghello et al [16] reported periarticular subchondral bone erosion adjacent to soft tissue calcification in a chronic renal dialysis patient. Steinbach et al [12] reported on tumoral calcinosis with bony erosion along the humeral head and femoral neck. Teng et al [17] reported erosive changes involving the posterior elements of the cervical spine in a patient with CREST syndrome. To our knowledge, cortical destruction with intramedullary involvement by tumoral calcinosis has not been reported

Tumoral calcinosis in patients with chronic renal failure commonly presents as large multilobular soft tissue masses with fluid-calcium levels. Although several cases of bone involvement have been reported, these represent bone erosion or remodeling rather than intraosseous penetration. To our knowledge, cortical destruction with intramedullary involvement by tumoral calcinosis has not been reported. Because of concern for pathologic fracture in the involved bone, radiologists should be aware of this rare complication of chronic renal disease.

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