Uncommon Masses in Renal Disease

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CASE ONE

A 26 year-old female with fever, tachycardia and pain in her left lower extremity, presented to the Emergency Department. Past medical history is remarkable for hypertension, end stage renal disease requiring hemodialysis and arteriovenous fistula (AVF). The AVF was performed in the left thigh undergoing superimposed infection. Physical examination revealed a fixed, non-erythematous tender mass at the left hip and an area of erythema, edema and induration at the surgical incision site of the arteriovenous access previously obtained at the left thigh. Blood chemistry values revealed hyperphosphatemia and normocalcemia. Computer Tomography (CT) of the left hip with contrast was ordered.

Figure 1: Computed Tomography of the left hip without contrast. (A) Coronal, (B) axial and (C) sagittal projections. Fluid levels present in both (B) and (C).

Figure 1A

Figure 1B

Figure 1C
CASE 2

A 38 year-old African American male with a past medical history of end stage renal disease due to hypertension since 1991, presented to our institution for a pre-transplant evaluation. The patient underwent a kidney transplant in 2001, however, due to non-compliance he developed progressive kidney rejection failure and has been on hemodyalisis since 2008. His blood chemistry laboratories values showed normal serum calcium levels. Chest radiographs at the time of examination were performed and compared with prior radiographs for further evaluation.

Figure 2: Chest radiographs. (A) Posteroanterior and (B) lateral views.

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RADIOLOGICAL DIAGNOSIS:
Tumoral Calcinosis with Fluid-Fluid Levels

IMAGING FINDINGS

In the first case, CT of the left hip with contrast revealed an amorphous calcified soft tissue mass with multicystic fluid levels located in the left proximal thigh Figure 1A, B and C (arrows) consistent with the clinical setting as above with tumoral calcinosis secondary to end stage renal disease. In the second case, prior chest radiographs demonstrated a small nonspecific ill-defined dense mass projected over the right upper chest wall. Follow-up radiographs seven years later revealed marked enlargement of the mass which appears to be lobulated and nodular with small fluid levels Figure 2 A and B (arrows). This was considered to be consistent with Tumoral Calcinosi s in the setting of long-standing renal disease.

DISCUSSION

Tumoral Calcinosi s is an uncommon familial disease secondary to a mutation in FGF23 (Fibroblast Growth Factor 23), KL (Klotho) gene and the UDP-Nacetyl-D-galactosamine:polypeptide N-acetylgalactosaminyltransferase 3 (GALNT3) gene. Tumoral Calcino sis was first described by Giard and Duret in 1898 and in 1943, Inclan et al named this entity as Tumoral Calcinosi s Characterized by densely, lobular calcified benign masses in soft tissue of the extensor surface of a bursa with characteristic imaging of amorphous, multilobulated cystic calcifications in the musculoskeletal system commonly periarticularly.

Currently, there are more than 300 cases of tumoral calcinosis have been described. It is most frequently seen in African Americans without sex predominance and usually appearing in childhood or young adulthood at the first and second decade of life. Symptomatic manifestation is uncommon unless there is compressive neuropathy. Large periarticular lesions can result in reduced range of motion. The most common locations of Tumoral Calcinosi s are the shoulder, hip, elbow, foot and wrist, but the temporoman diar joint, spine, hand and knee can also be involved.

The physical morbidity is based on repetitive trauma generating reparative impairment and initiation of osteoelastic activity. Smack et al, classified this entity into three types: (1) Primary normophosphatemic Tumoral Calcinosi s; in this type, patients have normal serum calcium and phosphate levels. (2) Primary hyperphosphatemic Tumoral Calcinosi s characterized by a defect in phosphate resorption and (3) Secondary Tumoral Calcinosi s: these patients have a concurrent disease that causes soft tissue calcification such as chronic renal failure with a secondary hypervitaminosis D, hyperparathyroidism and bone destruction. Imaging and biopsy may confirm the diagnosis revealing yellow material containing calcium hydroxyapatite.

Recent articles review the pathophysiology of Tumoral Calcinosi s in renal disease. Imaging studies include Radiographic examinations, Computer Tomography (CT), and Magnetic Resonance (MR) which can demonstrate dense nodular masses commonly with multiple fluid levels represented cystic structures with sedimentation of calcium which may communicate with the bursa of the adjacent joint. MRT1-weighted imaging shows inhomogenous lesions with low signal intensity; whereas on T2 weighted MR imaging, the lesion appears with a diffuse nodular pattern with areas of high intensity associated with signal low intensity or signal voids.

Other entities that can have similar imaging findings and that should be considered in the differential diagnosis of Tumoral Calcinosi s include: Calcinosi s Universalis, Calcinosi s Cirumscripta, Calcific Tendonitis, Synovial Osteochondromatosis, Synovial Sarcoma, Osteosarcoma, Myossitis Ossificans, Tophaceous Gout and Calcific Myonecrosis. In 1989, Kaplan showed a correlation between Tumorcal Calcinosi s and pseudoxanthoma elasticum and in 1990, Martinez et al described an association between Tumoral Calcinosi s and Calcific Myelitis.

Tumoral Calcinosi s treatment is based on its underlying cause, and is a combination between surgical excision and phosphate decrease secondary to acetazolamide administration. Acetazolamide increases calcium-phosphate solubility by lowering the systemic pH, resulting in an increase of phosphate excretion. Surgical excision is dependent on the patient’s symptoms due to compromise of the adjacent structures and further surgery for recurrence is recommended.

REFERENCES


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