

*Case reports*

# Imaging of tumoural calcinosis

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## Introduction

Tumoural calcinosis is an uncommon disorder characterized by the presence of calcified para-articular masses seen on radiographs. It was first described by Duret in 1899 [1,2]. It occurs at any age and is inherited as an autosomal dominant mode with variable clinical expression [1]. It is a relatively common clinical problem in Africa and occurs more commonly among black populations [3]. It is characterized by hyperphosphataemia and normocalcaemia with normal renal function.

## Case reports

In the past 2 years, four patients (two males aged 6 and 8 years and two females aged 13 and 21 years) with histopathologically-proven tumoural calcinosis presented to the surgical departments of King Hussein Medical Centre. The patients were not related.

The first patient, a 6-year-old boy, presented with right gluteal swelling which had been drained 1 year before because of a suspected abscess. Subsequently the patient developed a huge swelling in the right gluteal region which extended into the medial aspect of the upper thigh. Ultrasound scan was performed and showed a large cystic mass lesion with internal membrane and peripheral calcification (Figure 1)



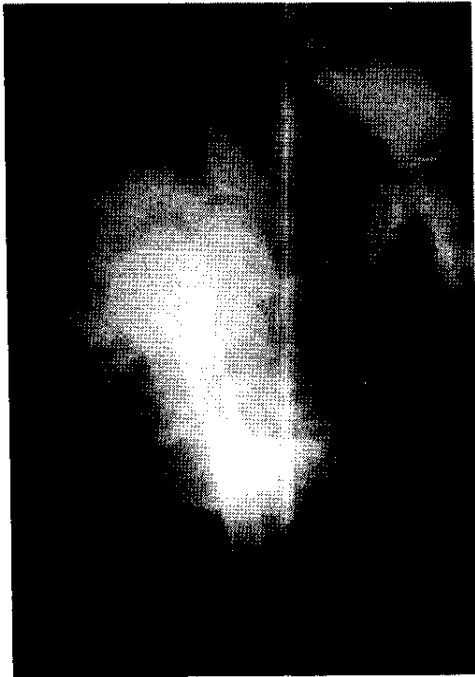
Figure 1 Ultrasound scans showing a large cystic lesion with internal membrane and peripheral calcification

which was misdiagnosed as complicated hydatid cyst. Plain film X-ray showed an inhomogeneous, lobulated, calcified soft tissue mass in the right gluteal region and upper thigh. The bones of the pelvis and right femur were normal (Figure 2). Computed tomography (CT) scan was performed and showed a large cystic mass posteriorly in the thigh with multiple low attenuation septation separating the nodular calcific component. Some of these septations showed dense calcium layering in the dependent part (sedimentation sign) (Figure 3).

Magnetic resonance imaging (MRI) showed a large nodular and hyperintense lesion alternating with areas of void signal

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**Figure 2 Right hip and femur showing inhomogeneous, lobulated, calcified soft tissue mass. The adjacent bones of the pelvis and femur are normal**



**Figure 3 Computed tomography scan showing a large cystic mass lesion with multiple low attenuation septations separating the nodular calcific components; some of them show dense calcium layering in the dependent part (sedimentation sign)**

on T2 weighted (TR = 4000 ms, TE = 90 ms) images (Figure 4). On T1-weighted images, the mass appeared inhomogeneous and had low signal intensity (Figure 5). Two other patients had similar lesions around the right hip; Figures 6 and 7 show CT and MRI images of one of these patients. The fourth patient, a 13-year-old female, had a similar lesion around the right elbow. Bone scintigraphy was not performed for our patients because diagnosis of tumoural calcinosis was made with other imaging techniques.



**Figure 4 T2-weighted coronal magnetic resonance imaging of the thigh showing a large mass with hyperintense nodules alternating with areas of void signal**

Surgical excision of the masses was performed for the four patients and all

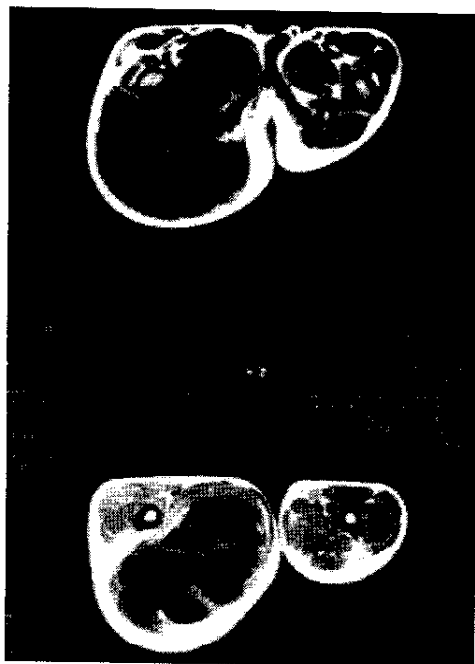


Figure 5 T1-weighted Images of the thigh showing an inhomogeneous large mass lesion with low signal intensity

proved to have tumoural calcinosis on histopathological examination.

## Discussion

Tumoural calcinosis is an uncommon syndrome of obscure etiology and pathogenesis. It is characterized by hyperphosphataemia, normocalcaemia and deposits of calcium salts in soft tissues, usually adjacent to large joints [1,2,4]. The lesions of tumoural calcinosis are divided into early and late stages [3]. Early stages consist of collagen necrosis and complete cyst formation. In addition, early stages may show unusual

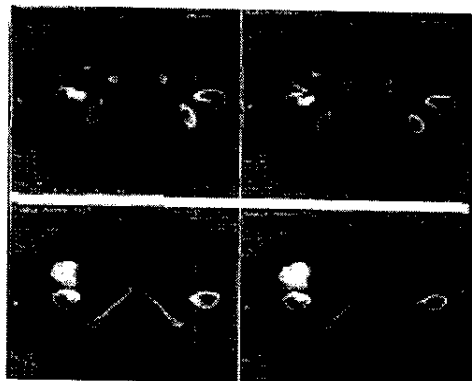


Figure 6 Computed tomography scan showing large cystic mass lesion with nodular calcific components located antero-medially; in addition, marrow calcification is noted in the proximal femur



Figure 7 Coronal magnetic resonance imaging for the same patient (Figure 6), showing decreased signal intensity within the marrow of the proximal femur due to marrow calcification

periosteal and marrow sclerosis which resolves over a period of several weeks without specific therapy [1,5]. Unfortunately our patients presented late and none

showed these early changes. In late stages, granular calcification occurs and is followed by dense calcification. In tumoural calcinosis the onset of soft tissue masses can range from age 3 months to 79 years, although they typically occur within the first and second decades of life [1].

Soft tissue masses are the best known component of the disease and characteristically occur in para-articular regions, most often in the hips, shoulders, elbows and feet. The masses tend to grow slowly over a period of years and may become quite large. Generally they are painless and do not limit the range of motion of adjacent joints, unless they become too large. They may result from compression of adjacent neural structures such as the sciatic nerve, and when large, tend to ulcerate the skin and form a sinus track that drains chalky milk-like fluid. This fluid may look like pus but is usually sterile and contains calcium phosphate and calcium carbonate [1,5,6,7]. The masses can, however, become a site of secondary infection.

Dental abnormalities, skin calcification and vascular calcifications are poorly recognized components of tumoural calcinosis, but none of these was observed in our patients. Specific biochemical abnormalities distinguish tumoural calcinosis as a unique metabolic bone disease, including hyperphosphataemia [1-3,5,7] and elevated serum vitamin D level. However, serum calcium, renal function, parathyroid hormone and alkaline phosphatase are normal [1,7]. Pathological features of the soft tissue masses are reflected in radiological findings. The mass is a granulomatous reaction to a foreign body, which may be active or inactive [1].

Imaging plays a major role in the investigation of tumoural calcinosis. The radiographic hallmark of tumoural calcinosis is the demonstration of large multiglobular

calcific deposits in a para-articular area, usually along the extensor surface of joints. The multinodular appearance is due to the radiolucent fibrous septa that separate the spaces containing the calcareous material (Figure 2). This calcified material may be paste-like and have a homogeneously dense radiographic appearance or it may be semi-fluid-like milk of calcium and demonstrate sedimentation on standing radiograph [1]. This sedimentation sign indicates a metabolically active lesion with a potential to grow in size or diminish in response to phosphate depletion therapy. The calcific deposit is usually localized to anatomic sites known to be occupied by bursae [1,3]. The bones adjacent to these calcific deposits are usually normal, but bone erosion may occur in rare instances. Para-articular calcareous masses are not exclusive to tumoural calcinosis, however. Similar masses can occur in patients with chronic renal failure who are undergoing haemodialysis or in patients known to have scleroderma [1,4].

CT imaging has been used to evaluate tumoural calcinosis. CT demonstrates the location of the masses deep in the buttock where the calcareous masses occupy the fibro-fatty plane deep in the gluteus maximus. The masses may extend proximally or distally in the facial planes or anteriorly and medially into the region of the sciatic notch. These masses often blend with adjacent muscle and give the appearance of intramuscular location or extension [6]. The masses may demonstrate the layering phenomenon believed to represent calcium salt in suspension [1] (Figure 3). The layering phenomena are indicative of a metabolically active lesion. CT, in addition, can image accurately marrow calcification (Figure 6).

MRI of para-articular masses of tumoural calcinosis has not been extensively studied. Calcific deposits, as expected, dis-

play long T1 and short T2 relaxation characteristics. Despite the known limitations of MRI for imaging calcific processes, other imaging techniques cannot image the inflammatory component in the para-articular masses. This can be best explained by the long T2 values associated with granulomatous foreign body reactions characteristic of tumoural calcinosis. In T2-weighted images, the lesion mass appears to contain hyperintense nodules, representing the inflammatory reaction involving the soft tissue, alternating with areas of void signal due to calcification (Figure 4). In T1-weighted images, the lesion mass appears inhomogeneous and of low signal intensity (Figure 5). MRI can show the marrow lesions properly; it appears as high signal intensity in T2-weighted images due to inflammation. Marrow calcifications will display decreased signal intensity in all MRI sequences (Figure 7). Small scattered foci of low signal intensity within the marrow in T1- and T2-weighted images are consistent with calcific deposits. The marrow changes seen in MRI are not specific and similar findings can be seen in neoplastic process, infarction, infection and trauma [1].

Radionuclide images have been used in diagnosing the para-articular soft tissue masses of tumoural calcinosis. It is the most reliable and simplest method for detection and localization and assessment of the calcific masses. Uptake of phosphate

compounds labelled with  $^{99}\text{Tc-m}$  by the calcific masses is probably related to surface absorption of hydroxyapatite crystal [1].

To our knowledge, the sonographic appearance of tumoural calcinosis has not been described before. TC masses on ultrasound appear cystic with an internal membrane or septation and show peripheral calcification. This may be mistaken for other soft tissue cystic masses, such as hydatid disease, especially in our country where echinococcosis is endemic (Figure 1). Ultrasound may also show layering of calcium salts in the dependent parts of the cyst, or the lesion may appear totally calcified.

In summary tumoural calcinosis is a rare hereditary metabolic disease characterized by a para-articular calcific process and associated with hyperphosphataemia but without other metabolic abnormalities. Imaging plays a major role in the diagnosis of tumoural calcinosis and can indicate the extent of the calcific masses. Plain radiography and CT are the best imaging techniques for diagnosis, however MRI plays a role in the assessment of activity of the lesion as well as its extension. Sonography has a limited role in the diagnosis of tumoural calcinosis; however, it can play a role in the diagnosis and follow-up of cases concomitant with plain films.

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