

Subscapular tumoral calcinosis in a toddler: case report

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ABSTRACT

Tumoral calcinosis is uncommon in toddlers, and rare within the subscapular area. Although typically benign, tumoral calcinosis is often incorrectly diagnosed prior to biopsy. We present a case of subscapular tumoral calcinosis in a 16-month old girl and discuss the radiological findings on X-ray, ultrasound, computed tomography and magnetic resonance imaging, including the first description of T1-weighted post contrast imaging, which demonstrate the fibrotic components of tumoral calcinosis.

CASE REPORT

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A 16 month-old girl presented with a large, painless, fluctuant mass along the left scapular border. Her past medical history was significant for prematurity of 24 weeks gestation, ligation of patent ductus arteriosus (PDA), retinopathy, necrotizing enterocolitis, nephrocalcinosis, and hypothyroidism.

A chest X-ray demonstrated a large, lobulated, heterogeneously calcified mass within the soft tissues of the lateral chest wall and deformity of the adjacent ribs (Fig. 1). Of note, however, rib deformities in the same area were observed on a previous post-operative radiograph from one year earlier, following ligation of a patent ductus arteriosus (PDA). Ultrasound showed a mixed solid and cystic mass (Fig. 2). Magnetic resonance imaging (MRI) demonstrated a large (7.5 x 4.3 x 3.2 cm) heterogeneous signal intensity mass within the soft tissues of the posterolateral chest wall deep to the left scapula extending intrathoracically through the intercostal spaces. The mass was isointense to muscle on T1-weighted (T1W) images, heterogeneously hyperintense on T2-weighted (T2W) images with multiple hypointense areas of calcification and fluid-fluid levels, and had heterogeneous post-contrast

enhancement (Fig. 3 A-C). Multiple internal non-enhancing cystic areas having fluid-fluid levels were observed. Computed tomography (CT) confirmed these findings, demonstrating extensive amorphous calcification of the mixed soft tissue mass within the intercostal spaces and extending into and outside the chest cavity, but not invading the pleural space (Fig. 4).

Serum calcium levels were slightly elevated (2.63 mmol per liter, normal range 2.10 - 2.55), phosphate levels were normal (1.75 mmol per liter, normal range 1.10 - 2.10) and parathyroid hormone levels were low (8 ng per liter, normal range 13 - 54). Based on the radiological findings, differential diagnoses included: post-traumatic calcified hemangioma or lymphangioma resulting from previous PDA ligation; hamartoma; teratoma; or a soft tissue sarcoma.

Biopsy demonstrated tumoral calcinosis (TC) as characterized by dense fibrous connective tissue surrounding calcified areas bordered by multinucleated giant cells and histiocytes (Fig. 5).

The mass was excised under general anesthesia with no complications (Fig. 6). During the surgery, chalky appearing

fluid extruded from pseudocapsules, consistent with previously described cases of TC. Aside from a large seroma, which resolved spontaneously two weeks post-operatively, the patient remained well with no recurrence of the mass at two months following its excision.

DISCUSSION

Tumoral calcinosis (TC), first described in 1898 [1], is characterized by the development of one or more soft tissue masses with significant deposition of amorphous calcium salts, typically occurring around large joints, commonly hips, knees, shoulders and elbows [2,3]. Its incidence is unknown; most reported cases have occurred in Black adolescents or young adults of African descent [1,2].

The etiology of TC remains unclear, but can be classified into three groups resulting from: (1) a genetic condition, an autosomal recessive mutation in one of three genes (fibroblast growth factor (FGF) 23, KL, and GALTN3) which induces errors in phosphate metabolism [4], (2) a complication of renal failure and dialysis, or (3) a sporadic occurrence [5]. Many sporadic cases have been linked to previous trauma. [6]

While hyperphosphatemia is common in TC, serum levels of calcium and parathyroid hormone (PTH) tend to be normal [7]. The mutations in FGF23 and GALTN3 are known to increase the reabsorption of phosphate in the proximal tubule, increasing serum phosphate levels. This, in turn, could be expected to cause hypocalcemia; however, Janssen et al (2009) theorized that this is counterbalanced by elevated levels of calcitriol, which would stimulate increased reabsorption of calcium in the distal nephron [7]. The combination of excess phosphate and normocalcemia often causes calcium and phosphate to deposit in the skin and subcutaneous tissues [7]. Interestingly, TC has been described as the metabolic 'mirror image' of rickets, which is often caused by upregulation of FGF23 by either mutation or a tumour, and results in decreased phosphate reabsorption in the nephron [8].

Clinical findings vary among patients: some masses are painful and swollen while others are not [8]. Many of the masses cause erosion of the skin and extrude a white or yellow chalky substance resembling pus. [6]

The case presented here is particularly rare in terms of both the anatomical location of the TC in a subscapular location, and the age of the patient. A single analogous case was recently reported in the literature. Four months following repair of coarctation of the aorta, a six-month old girl developed TC within the thoracotomy scar, displacing the left scapula [5]. The tumour was resected but recurred twice more. The third resection at the age of 22 months was thorough, and there were no signs of recurrence 12 months later [5].

One additional case of tumoral calcinosis in the scapular area was described in a 9 year-old girl with a mass below the inferior border of the left scapula [8]. No previous trauma was reported in this patient, and serum calcium and phosphate levels were normal [8].

Other than these two patients, only 16 other cases of TC in infants or toddlers have been reported in the literature. In contrast to TC cases in older patients, no predisposing factors (trauma, metabolic, or familial) could be identified in most infantile cases [9], suggesting a different etiology. The masses tended to occur in different locations than in adults such as wrist, ankles, toes, and in the supraclavicular area [9], possibly due to these joints being used more actively in this age group.

Our patient presented with a large mass displacing the left scapula, in the region of a previous surgery. Radiography, CT and MRI results were consistent with previously described cases of TC, which have similar findings in adults, children, and infants [8,10]. On all imaging modalities, TC displays a cobble-stone, or 'chicken-wire' appearance representing amorphous calcium deposits in grape-like clusters and separated by fibrous bands [10]. Dense calcium layering (fluid-fluid levels) in the dependent part, known as the sedimentation sign, can be observed in some patients but is non-specific to TC [8,10]. It was noted in our patient.

While CT is best for demonstrating the extensive calcification within TC masses, MRI may be useful in depicting the tissue characteristics of TC. These masses are typically hypointense on T1W and heterogeneously hyperintense on T2W images, sometimes with fluid-fluid levels apparent [8]. These reflect the absence of fat, and high water content or inflammation, respectively. T1W post contrast imaging in TC has not previously been reported. Our patient demonstrated heterogeneous hyperintense areas, likely reflecting the fibrous components of TC.

The etiology in our patient remains uncertain due to a complex history including significant prematurity, nephrocalcinosis, slightly elevated serum calcium levels, hypothyroidism, and previous surgery in the area of the mass. The presence of normal phosphate levels suggests that this is not a familial case of TC. Based on previous reported cases of TC, it seems the most likely etiology is operative trauma from a previous PDA ligation; however a metabolic cause or sporadic occurrence cannot be excluded.

Tumoral calcinosis is often not considered as a potential diagnosis in very young patients, and has been depicted as difficult to diagnose even in adult patients. Despite distinctive radiological features, Steinbach et al observed that five of twelve patients were misdiagnosed prior to biopsy, and their initial diagnoses included osteosarcoma, chondrosarcoma, abscess, and a herniated bowel [10].

In addition to tumoral calcinosis, which is rare in toddlers, the differential diagnosis for a calcified soft tissue mass is lengthy and includes: heterotopic ossification (myositis ossificans), calcified hemangioma or lymphangioma, teratoma, parosteal osteosarcoma and soft tissue sarcomas, metachondromatosis, chronic renal failure, calcinosis universalis, hypervitaminosis D, milk-alkali syndrome, and calcinosis circumscripta. TC has a classic appearance of a lobulated calcified mass in a periarticular location with no involvement of the adjacent bones. Heterotopic ossification is comprised of heterotopic bone and cartilage typically located

within muscles. It may be differentiated from TC by its rapid evolution from faint calcification to heterotopic ossification and by a lack of lobular morphology. Calcified hemangioma is seen as an intensely enhancing soft tissue mass with multiple phleboliths. A lymphangioma is typically a multicystic nonenhancing mass and extensive calcification is uncommon. A teratoma specifically contains areas of fat attenuation, whereas TC does not contain fat. Parosteal osteosarcoma is also seen as an ossified soft tissue mass, however is nearly always connected to the underlying bone through a stalk. Soft tissue sarcomas are aggressive and infiltrative in nature and may have areas of calcification. The underlying bones may be involved depending on the severity of the mass. Metachondromatosis, an inherited skeletal disorder, which leads to multiple enchondromas and osteochondromas and does not involve the soft tissue.

Calcinosis universalis is associated with collagen vascular diseases and is recognizable by its sheet-like deposits of calcium within the muscle, subcutaneous tissue and the fascia. Calcinosis circumscripta can appear similar to TC, but is less extensive and occurs in the subcutaneous tissues as focal, dense, and well-defined calcifications.

Finally, milk-alkali syndrome can result from ingestion of too much calcium, thus raising the serum calcium levels and causing deposits of calcium, particularly in the kidneys (nephrocalcinosis). Hypervitaminosis D, in effect, causes hypercalcemia which may also lead to calcium deposits. Chronic renal failure can impair phosphate reabsorption, and thus is a cause of secondary TC, having similar pathophysiology to familial TC and is indistinguishable in its radiological and histological features.

Most TC masses in infants or toddlers were removed surgically without recurrence [9]. However, in adults and cases where a predisposing genetic factor was identified, recurrence is common [6,9]. Phosphate deprivation and use of acetazolamide are often prescribed to decrease rates of recurrence in adults [6,9]. Reports of spontaneous regression of TC without any treatment have also been reported [11].

TEACHING POINT

While tumoral calcinosis is rare, it can present at any age in nearly any location and should remain in the differential diagnosis for any calcified soft tissue mass. Its various components of calcium deposition, fibrous bands and soft tissue contribute to distinctive radiological features including extensive calcification without a connection to bone, well visualized on CT, a cobble-stone appearance with or without the sedimentation sign, and iso- or hypointensity on T1W, and heterogeneous hyperintensity on both T2W and T1W post contrast MRI images.

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FIGURES



Figure 1. 16 month-old female with subscapular tumoral calcinosis. PA radiograph showing lobulated, calcified mass along the left chest wall measuring 8.5 by 3 cm (arrow) and left rib deformities (arrowhead) secondary to previous PDA ligation.

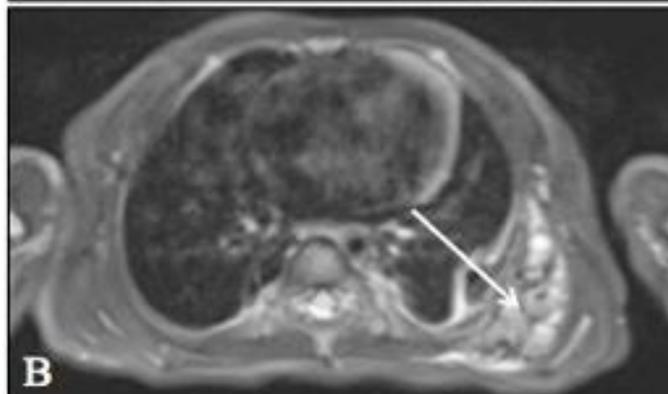
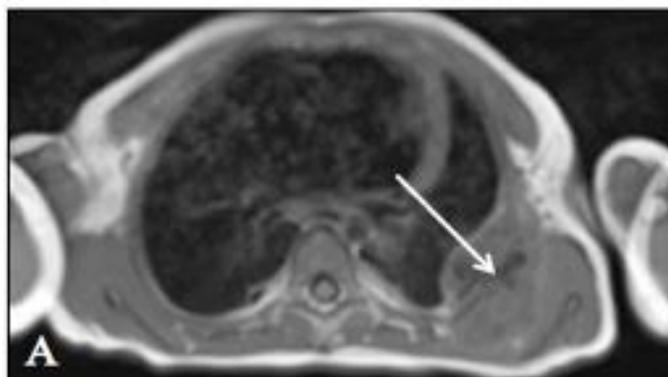


Figure 3. 16 month-old female with subscapular tumoral calcinosis. Axial MRI images show a heterogeneous mass in the left subscapular region appearing (A) isointense to muscle on T1 (TR: 783 ms, TE: 14 ms, slice thickness: 5 mm, spacing 5.5 mm) and (B) heterogeneously hyperintense on T2 (TR: 3500 ms, TE: 67 ms, slice thickness: 5mm, spacing 5.5 mm) with fluid-fluid levels and low intensity areas, suggestive of calcification. (C) Coronal T1 post contrast image shows heterogeneous enhancement of the mass. (TR: 620 ms, TE: 14 ms, slice thickness: 3 mm, spacing 3.3 mm.) The mass, approximately 7.5 cm in the craniocaudal dimension and 4.3 x 3.2 cm in axial transverse dimensions, has intrathoracic extension through the intercostal spaces with extrapleural involvement, but no pulmonary involvement. Siemens Avanto system, 1.5 Tesla; 1.7 mL Magnevist.

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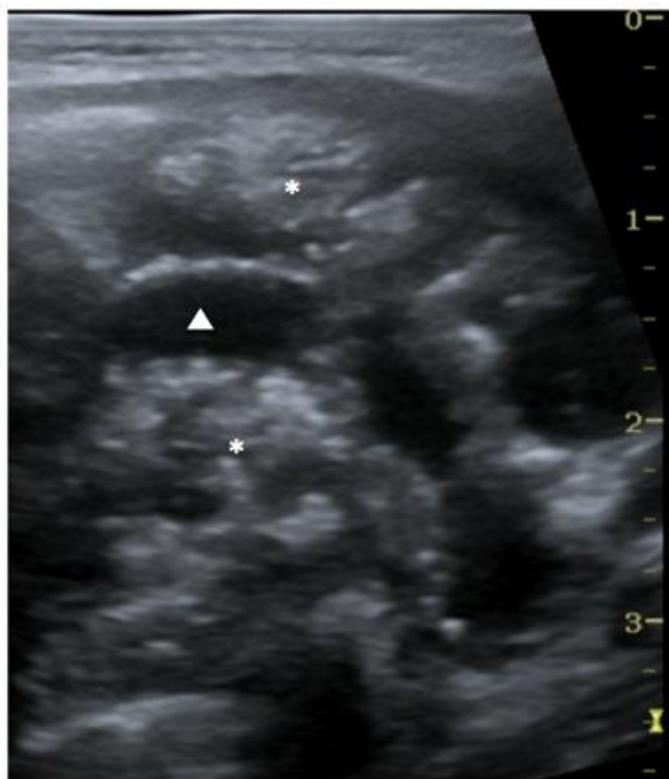


Figure 2. 16 month-old female with subscapular tumoral calcinosis. Sagittal ultrasound image showing heterogeneous mixed solid and cystic mass (asterisks), measuring approximately 4.5 x 2.7 x 3.7 cm in size, within the soft tissues below the left scapular border and adjacent to a rib (triangle). 9 MHz linear transducer, depth: 4.0 cm.

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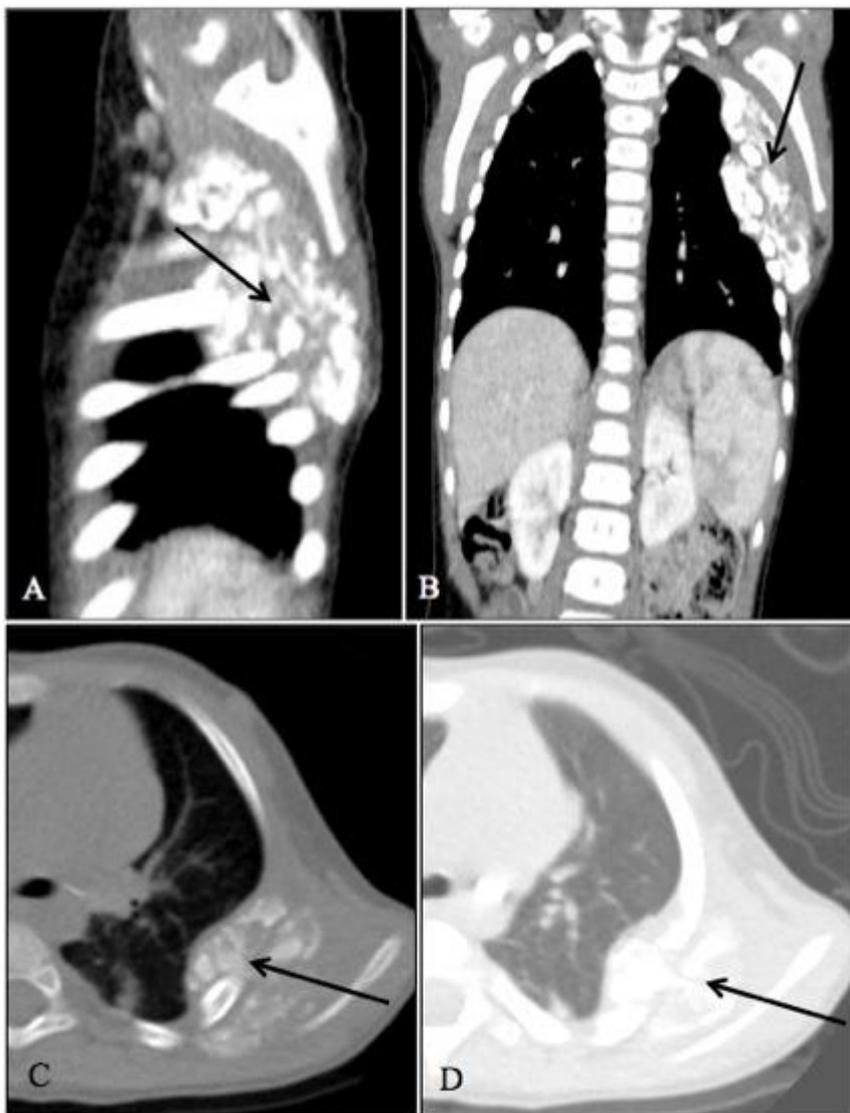
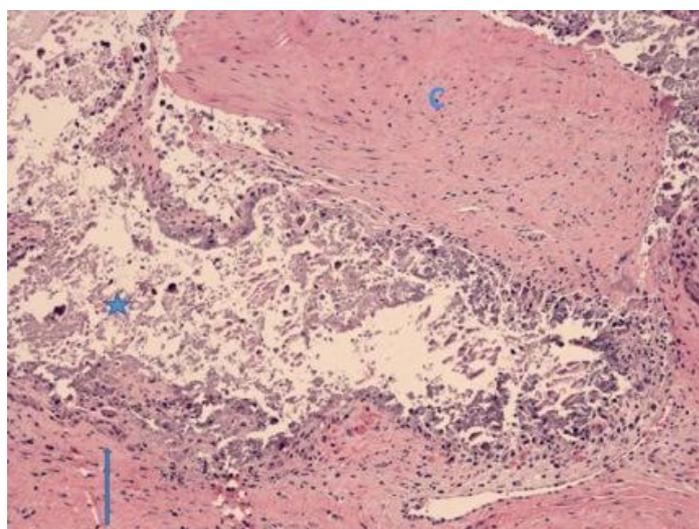


Figure 4 (left). 16 month-old female with subscapular tumoral calcinosis. (A) Sagittal, (B) coronal, and (C, D) axial CT reformat image showing dense amorphous calcification within the left subscapular mass, measuring 7.5 cm in craniocaudal dimension and 4.1 x 3.2 in biaxial transverse dimensions. (D) demonstrates mass in lung window. GE LightSpeed VTC, 64 slice scanner, 100 kV, 92 mA, slice thickness: 2.5 mm, contrast: 16 ml Optiray 320, given by hand.

Figure 5 (right). 16 month-old female with subscapular tumoral calcinosis. Photomicrograph demonstrating granular deposits of calcified material (star), bordered by histiocytes with multinucleated giant cells (arrow) and separated by dense bands of collagenous connective tissue (C). Hematoxylin and eosin. Original magnification x100.



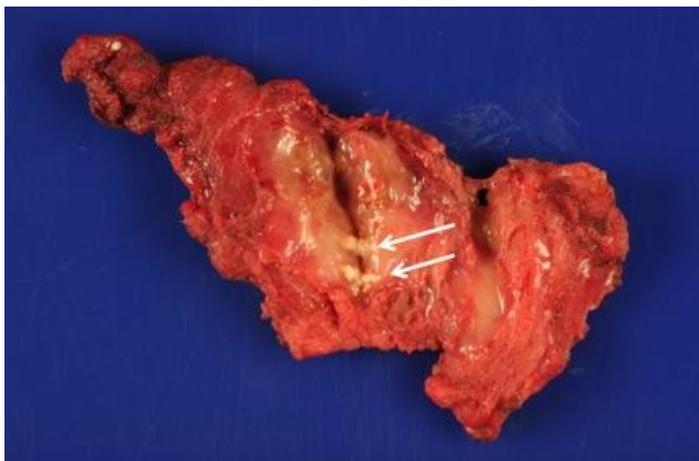


Figure 6 (left). 16 month-old female with subscapular tumoral calcinosis. Resected specimen (measuring 4.0 x 3.0 x 0.7 cm) showing small yellow areas of calcification (arrow). The mass was resected in two fragments; the larger fragment is shown in this figure. The smaller fragment (not shown) measured 1.8 x 1.4 x 0.5 cm.

Etiology	<ol style="list-style-type: none"> 1) Complication of renal failure/dialysis 2) Genetic disorder affecting phosphate metabolism 3) Sporadic/unknown; possibly linked with trauma
Incidence	Not reported – only 18 cases in infants described in the literature
Gender ratio	Not reported
Age predilection	Adolescents and young adults
Risk factors	African descent, genetic phosphate metabolism abnormalities
Treatment	Surgery, but recurrence is possible. Phosphate restriction and acetazolamide may prevent recurrence. Spontaneous regression can also occur.
Prognosis	Good. Masses are benign and often successfully removed without recurrence if there are no predisposing factors.
Findings on imaging	<ul style="list-style-type: none"> • General: a cobble-stone appearance with or without the sedimentation sign • X-ray: calcified mass • Ultrasound: mass demonstrating mixed solid and cystic components • CT: dense amorphous calcification • MRI: <ul style="list-style-type: none"> ○ T1W: hypo- or isointense ○ T2W: heterogeneously hyperintense ○ Post contrast: heterogeneously hyperintense

Table 1: Summary table for tumoral calcinosis

	CT	MR	X-Ray
Tumoral calcinosis	Amorphous, cystic, multilobulated calcified mass in periarticular location. Sedimentation sign may be present. Adjacent bones are normal.	Hypo or isointense on T1W; heterogeneously hyperintense with hypointense areas of calcification on T2W. Post contrast: heterogeneously enhancing.	Multilobulated rounded calcified soft tissue mass in periarticular location usually affecting extensor surfaces.
Calcified Hemangioma	Intensely enhancing mass with multiple phleboliths/calcification.	Seen as hypointense mass on T1 and diffusely hyperintense on T2 with intense enhancement.	Multiple small phleboliths within a soft tissue mass.
Calcified Lymphangioma	Soft tissue mass of multiple homogeneous nonenhancing areas with variable attenuation values. Calcification is uncommon.	Multicystic lesion, hypointense on T1, hyperintense on T2 with relatively no enhancement or peripheral enhancement of cystic spaces. Calcification is uncommon but may be seen as hypointense areas on gradient images.	Soft tissue mass with faint concentric areas of calcification. Difficult to appreciate on radiographs.
Parosteal osteosarcoma	Densely ossified mass arising from cortical surface of the bone.	Extraskeletal ossified mass arising from the surface of bone seen as hypointense on T1 and T2 with heterogeneous post contrast enhancement.	Densely ossified mass originating from surface of the bone. If connecting stalk not visible, mimics tumoral calcinosis.
Teratoma	Mixed attenuation soft tissue mass with solid and cystic areas. Areas of fat attenuation and calcification.	Heterogeneous signal intensity mass lesion with T1 hyperintense fatty component and T2 hypointense areas of calcification.	Soft tissue mass with variable calcific and fat densities.
Soft tissue sarcomas	Aggressive and infiltrative in nature and may have areas of calcification. The underlying bones may be involved depending on the severity of the mass.	Poorly defined infiltrative soft tissue mass with solid and cystic areas seen as heterogeneous signal intensity on T2 along with heterogeneous enhancement. Multiple nonenhancing areas of necrosis may be seen. Calcification may be seen in part of the tumor.	Lobulated dense calcific areas seen within a part of the soft tissue mass. Erosion or destruction of the underlying bone.
Metachondromatosis	Bone lesion with multiple osteochondromas and enchondromas. Hands and feet and long bones and iliac crests. Exostoses point towards the joints. No soft tissue component.	Lobulated lesions with intermediate signal on T1 and predominantly high signal on T2.	Multiple exostoses develop inside as well as outside of the bones, slope of the exostoses towards epiphysis.
Chronic renal failure	Periarticular calcified mass. No radiologic differences from tumoral calcinosis (cause of secondary tumoral calcinosis). Vascular calcifications may also be present.	T1 and T2 hypointense calcifications.	Periarticular calcified mass indistinguishable from tumoral calcinosis. Diagnosis by history and serum chemistry.
Calcinosis universalis	Diffuse, sheet-like deposition of calcium involving muscles, subcutaneous tissues and fascial planes.	T1 and T2 hypointense for calcifications in sheet-like configurations. Possible muscular or subcutaneous increased signal intensity on STIR (representing edema).	Widespread linear and sheet like soft tissue calcification along the muscular and fascial planes. Association with connective tissue diseases.
Hypervitaminosis D	Periarticular soft tissue calcifications. Finely speckled calcifications.	Hypointense on T1- and T2-weighted images.	Speckled periarticular calcifications, clinical history, hypercalcemia and hypercalciuria distinguish from tumoral calcinosis.
Milk-alkali syndrome	Periarticular soft tissue calcifications. Finely speckled calcifications.	Hypointense on T1- and T2-weighted images.	Speckled periarticular calcifications associated with hypercalcemia and metabolic alkalosis, associated nephrocalcinosis.
Calcinosis circumscripta	Less extensive soft tissue calcification than tumoral calcinosis. Located in subcutaneous tissues rather than bursal regions. Focal, dense, well-defined calcifications.	Hypointense on T1- and T2-weighted images.	Nodular calcification in subcutaneous tissues less extensive than tumoral calcinosis. Associated with connective tissue diseases.

Table 2: Differential diagnosis table for tumoral calcinosis

ABBREVIATIONS

CT = computed tomography
MRI = magnetic resonance imaging
PDA = patent ductus arteriosus
T1W = T1-weighted
T2W = T2-weighted
TC = tumoral calcinosis

KEYWORDS

Tumoral Calcinosis; Magnetic Resonance Imaging; Computed Tomography; Case Report

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