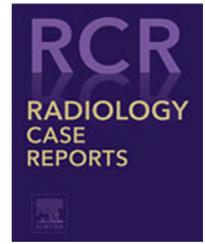


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## Case Report

# Hyperphosphatemic familial tumoral calcinosis mimicking a cystic hemo-lymphangioma on MRI <sup>☆</sup>

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## ABSTRACT

A tumoral calcinosis is a rare benign pathology characterized by calcium deposits (calcium phosphate crystals) in the periarticular soft tissues, giving a truly pseudotumor appearance. The same patients with tumoral calcinosis may have manifestation of hyperostosis hyperphosphatemia syndrome. The association is called Hyperphosphatemic familial tumoral calcinosis which is the case with our patient. We present a unique case of a 10-year-old female child without any notable history. No notion of consanguinity, a non-painful swelling of the right elbow for the last 3 years. She was presented with tumoral calcinosis in the context of familial hyperphosphatemic calcinosis tumor in which the diagnosis of lymphangioma was evoked and then redressed.

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## Introduction

A tumoral calcinosis is a rare benign pathology characterized by calcium deposits (calcium phosphate crystals) in the periarticular soft tissues, giving a truly pseudotumor appearance, hence its name. Two forms are distinguished: the primary form of probable genetic origin, also called familial, and the secondary form.

The presentation on magnetic resonance imaging of pseudotumor calcinosis can be misleading and simulating a cal-

cified cystic hemangio-lymphangioma. We report the case of a 10-year-old girl who was presented with a swelling of the right elbow that was initially diagnosed as a cystic hemo-lymphangioma. The final diagnosis retained was a hyperphosphatemic familial tumoral calcinosis.

## Case report

This is a 10-year-old female child without any notable history, no notion of consanguinity and a non-painful swelling of the right elbow for the last 3 years.

<sup>☆</sup> Competing Interests: None.

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**Fig. 1 – The appearance on standard radiography is a multilobular para-articular calcified lesion giving an cloud-like appearance.**

The clinical examination found a mass opposite the elbow; it revealed a hard mass of the soft parts, not painful to palpation and without any skin signs. An initial MRI (Fig. 5) was performed which revealed a multiloculated lesion formation in leger T1 hypersignal and T2 hypersignal. Some of the cells having several liquid levels, on the T2-weighted sequence (hyposignal - intermediate signal and hypersignal indicating a shading sign) (Fig. 5A) related to probable hemorrhages of different ages, with intense enhancement of the walls of the cystic cells after injection of gadolinium (Fig. 5B). In the light of these MRI data, the diagnosis of cystic hemolymphangioma was evoked.



**Fig. 2 – Ultrasound the appearance is that of a multilocular lesion with cystic pockets and attenuating calcification.**

A biopsy of the mass was performed and came back in favor of a calcinosis (Fig. 6). In view of the radio-histological discrepancy and the need to look for calcifications that were not well analyzed on the MRI scan, a triplet of standard radiography, ultrasound and CT scan was performed and the data collected were as follow:

- ✓ Standard radiography (Fig. 1) of the elbow revealed a voluminous calcified pseudotumor mass of the soft tissues in front of the elbow, poly-lobed, with regular contours without signs of bone invasion respecting the humeral cortical and the elbow joint.
- ✓ Ultrasound (Fig. 2) of the elbow mass was in favor of several thin-walled cavities containing pure and thick fluid with the presence of several attenuations.
- ✓ The CT (Fig. 3) scan with injection of the contrast product shows a mass of soft parts in front of the elbow, multiloculated, with regular contours and individualization of calcic levels within the cystic stalls and parietal enhancement. After injection of the contrast product, absence of periosteal reaction and absence of articular invasion.

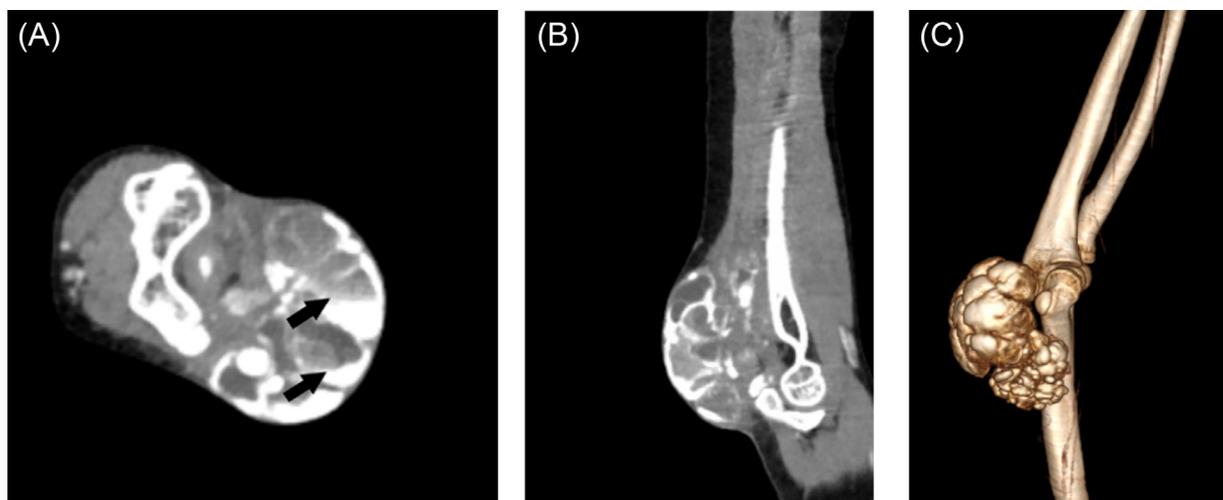
In view of the new radiological data, the diagnosis of primary pseudotumor(or tumoral) calcinosis was evoked certainty and reinforced by the histological data.

The complement of biological assessment is reviewed with hyperphosphatemia and a normal renal function.

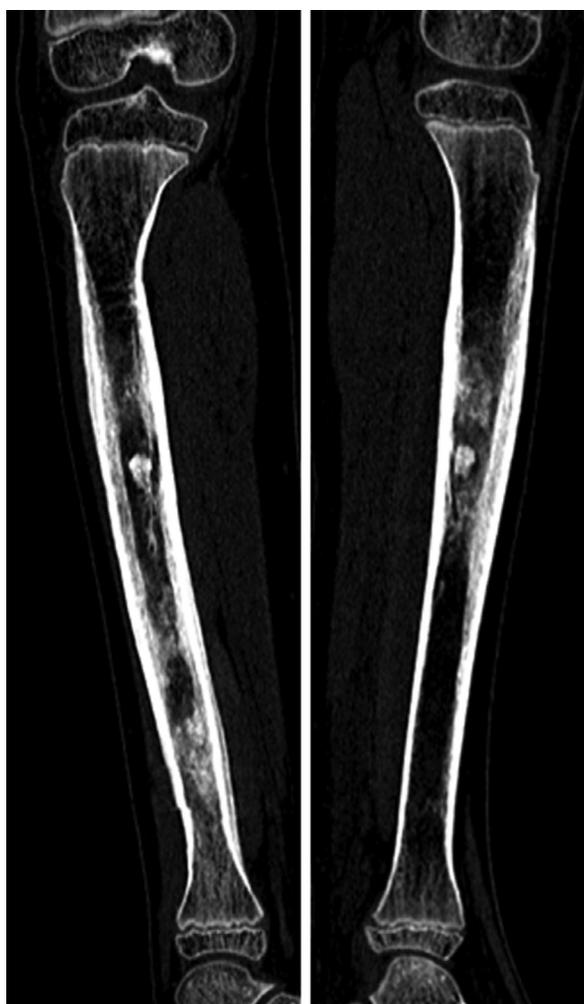
In addition, the patient had pain in both legs. A CT scan was performed showing diaphyseal sclerosis and mild periosteal reaction of the tibia related to diaphysitis within the framework of hyperostosis hyperphosphatemia syndrome (HHS) (Fig. 4).

The final retained diagnosis was a hyperphosphatemic familial tumoral calcinosis (association tumoral calcinosis and HHS).

The mass was totally resected following the cleavage planes between it and the muscular planes. The piece was sent to the histological analysis.



**Fig. 3** – CT acquisition in axial section (Figure 3A), in sagittal reconstruction (Figure 3B) and 3D (Figure 3C) shows a multilocular lesion with cystic pockets containing a level reflecting the sedimentation of calcium crystals in a serous supernatant (figure 3A , black arrows).



**Fig. 4** – CT coronal acquisition show a hyperostosis with cortical thickened.

Follow-up in internal medicine was proposed to the patient.

## Discussion

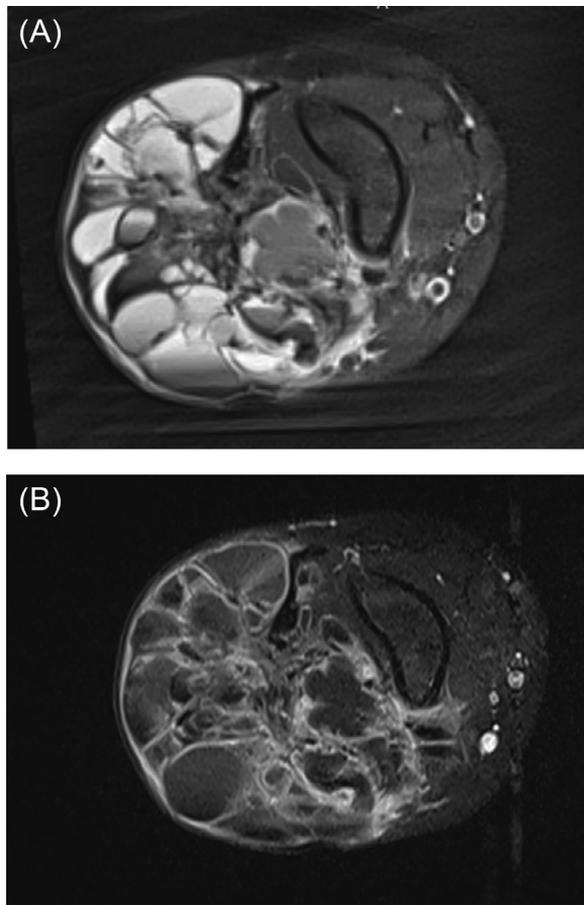
Tumoral calcinosis is a rare benign tumor or pseudotumor whose primary forms have not been frequently reported in the literature [1].

Described for the first time by Giard in 1898, it was named by Teutschlaenderqui—lipocalcinogranulomatosis—and defined as a fatty necrosis and an accumulation of calcium carbonate. In 1943, Inclan was the first to use the term tumor calcinosis [2].

The most prevalent locations of tumoral calcinosis in decreasing order are hip, elbow, shoulder, foot, and wrist [3].

There are 2 forms: the secondary form, which is generally a consequence of a disorder of phosphocalcic metabolism (renal insufficiency, hyperparathyroidism, and certain systemic diseases) and the primary form, also called familial, the mechanism of which is not yet well understood, but which probably has a genetic origin with an autosomal recessive transmission [4]. This will be responsible for an enzymatic disturbance causing familial hyperphosphatemia, which coinciding with normal renal the case with our patient. Several factors (trauma and microtrauma) may explain the mechanism of occurrence of pseudotumor calcinosis in patients without secondary causes or family antecedents.

The appearance of pseudotumoral calcinosis on standard radiography is a multilocular para-articular calcified lesion giving an cloud-like appearance. CT shows a multilocular lesion with cystic pockets containing a level reflecting the sedimentation of calcium and MRI sequences shows a multiloculated lesion formation in leger T1 hypersignal and T2 hypersignal, with some of the cells having several liquid levels with different signals on the T2-weighted sequence and intense en-



**Fig. 5 – MRI sequences shows a multiloculated lesion formation in leger T1 hypersignal and T2 hypersignal, with some of the cells having several liquid levels with different signals on the T2-weighted sequence (FIGURE 5A), with intense enhancement of the walls of the cystic cells after injection of gadolinium (FIGURE 5B).**

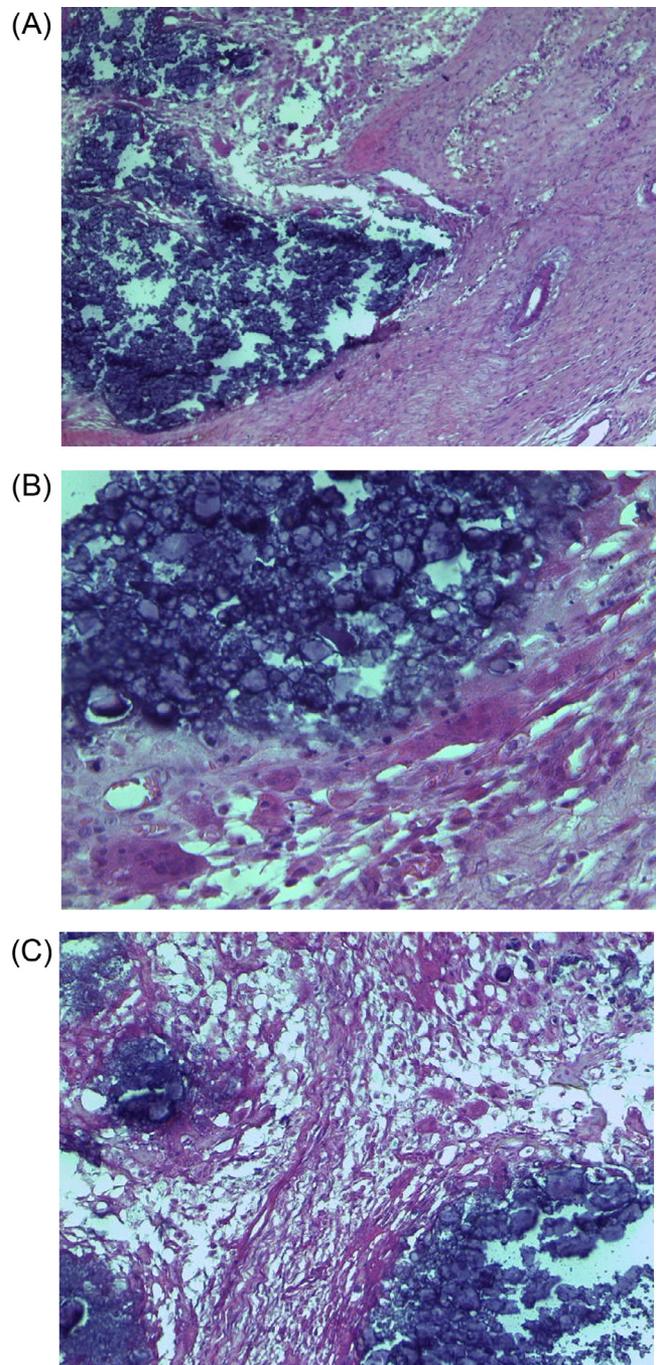
hancement of the walls of the cystic cells after injection of gadolinium.

At histopathologic examination, epithelioid elements and multinucleated giant cells surround calcium granules [3].

Hyperostosis hyperphosphatemia syndrome (HHS) is characterized by involvement of the long bones associated with elevated serum phosphate secondary and a mutation affecting either FGF23, caused decreased FGF23 signaling. This syndrome occurs by painful swellings in the areas overlying the diaphysis of the tibia. Imaging showed medullary sclerosis and extensive periosteal new bone [5].

Same patients with tumoral calcinosis may have also manifestations of HHS, the association is called Hyperphosphatemic familial tumoral calcinosis, which is the case with our patient.

Lymphangiomas are a diverse group of lymphatic channel vascular malformations made up of cystically dilated lymphatics. These benign abnormalities, according to Landing and Farber [6], can be divided into 4 groups: capillary lymphangioma, cavernous lymphangioma, cystic lymphangioma (hy-



**Fig. 6 – Microscopic examination shows large calcific foci arranged in lobules and separated by fibrous septa harboring a polymorphic inflammatory infiltrate and an inflammatory reaction rich in histiocytes and foreign body giant cells. Figure 6 A : Calcifications within a fibro-vascular tissue, figure 6B : calcinosis with foreign body reaction and presence of giant cells around the calcifications, figure 6C : Calcifications of varying size.**

groma), and hemo-lymphangioma (combination of heman-gioma and lymphangioma). The hemo-lymphangiomas seems to develop from circulatory system malformations that are present at birth. The blockage of venolymphatic communica-

tion between dysplastic vascular tissue and the systemic circulation may have contributed to the development of that tumor.

Standard radiography, echography, and CT show a typical feature of the calcinosis pseudotumor like a voluminous calcified pseudotumor mass of the soft tissues in front of the joint with individualization of calcic levels within the cystic stalls and parietal enhancement. The calcic levels may mimicking hemorrhages levels on MRI and wrongly evoking the diagnosis of hemo-lymphangiomas. So on MRI, it is challenging to discern between the calcific component of pseudotumor calcinosis and the hemorrhagic content of this lymphatic tumor; however, the standard radiography and the CT find all their interest.

There are several diagnoses having a similar aspect in standard radiography and CT because of calcifications. Many known examples such as dermatopolymyositis, calcific tendonitis, calcific bursitis osteosarcoma [3].

The diagnosis of calcinosis pseudotumoris is easy to get on standard radiography and CT scan; it doesn't usually require MRI, which was not done in our patient for whom it was requested in the first instance.

The interest of this article is to unveil the most common error which starts with an MRI analysis and shunt a standard radiographic or ultrasound to diagnose soft tissue disease. Also, to present manifestation of hyperphosphatemic familial tumoral calcinosis which is a rare benign disease.

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## Conclusion

Hyperphosphatemic familial tumoral calcinosis, associate a manifestation of tumoral calcinosis and hyperostosis hyperphosphatemia syndrome (HHS). A calcified mass in front of joint and diaphysitis and also hyperphosphatemia.

The standard radiography and the CT find all their interest in this disease to detect calcifications and not to miss the diagnosis.

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## Patient consent

The authors declare that the patient consents for publication of their case.

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