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# Tumoral calcinosis of the foot with unusual presentation in an 11-year-old boy: A case report and review of literature

Mohamed S, Jong-Hun J, Weon-Yoo K

Department of Orthopedics,  
Catholic University, Daejeon  
St. Mary's Hospital, South  
Korea

## Correspondence:

Mohamed Shafi  
E-mail: drmohamedshafi@  
hotmail.com

## ABSTRACT

Tumoral calcinosis is an uncommon disorder characterized by development of calcified masses within the soft tissues near the large joints such as the hip, elbow and shoulder and rarely occurs in the foot. We report the clinical, radiological and magnetic resonance imaging findings in a case of tumoral calcinosis of the foot observed in an 11-year-old boy which caused erosion of underlying bone and describe the surgical treatment and pathology results.

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Tumoral calcinosis is an unusual benign condition characterized by large calcified periarticular soft tissue masses composed of calcium salts, usually located around large joints. There is a primary form (idiopathic or hereditary) but tumoral calcinosis can also be found in a wide variety of conditions secondary to hyperparathyroidism, vitamin D intoxication, scleroderma and uremic tumoral calcinosis.<sup>[1]</sup> Calcification essentially limited to the soft tissues with only the occasional pressure erosion of underlying bony surfaces has been reported.<sup>[2]</sup> We report a case of tumoral calcinosis at the distal inter-phalangeal joint (DIP) of the fifth toe in an 11-year-old boy after a minor injury, which caused erosion of underlying bone which was excised surgically in order to prevent the mal-growth of the bone. There were unusual findings, such as normal calcium and phosphate levels, as well as peculiar presentation of the pathology and the implications about its treatment are worthy of description.

## Case History

An 11-year-old boy visited our hospital with complaints of pain and swelling in the DIP of the fifth toe. This pain had occurred for three weeks after a minor injury to this area. There was no family history of a similar complaint and the patient was otherwise healthy. On physical examination he had a soft, well-defined 1 x 1 cm mass over the medial aspect of the DIP of the fifth toe and the area was sensitive to the touch. There was minimal surrounding erythema and no drainage

or skin breakthrough. The laboratory examination showed normal calcium (9.0 mg/dl), phosphorus (3.9 mg/dl), serum alkaline phosphatase (35 IU/L) and uric acid (3.5 mg/dl). An antero-posterior (AP) radiograph revealed a well-defined, multiloculated, 1x1.2 cm calcified juxta-articular mass in the medial aspect of the DIP of the fifth toe [Figure 1]. This was associated with erosion of the underlying bone and there was soft tissue swelling as well. An MRI of the foot was done to assess the extent and anatomical position of the para-articular mass. T1 and T2-weighted images showed a soft tissue mass



Figure 1: AP radiograph of the right foot shows a well-defined, multiloculated juxta-articular calcified mass in the medial aspect of the fifth DIP joint

including multiple dark signal intensity nodules and peripheral intermediate signal intensity in the juxta-articular area of the DIP joint of the fifth toe [Figure 2]. On a post contrast T1-weighted image, a moderate enhancement was seen in the intermediate signal intensity lesion.

The patient underwent an excision biopsy and curettage under spinal anesthesia. The skin incision was made on the plantar aspect of the fifth toe from the proximal inter-phalangeal joint (PIP) to the DIP joint. After meticulous dissection, a chalky white material measuring 1x 2 cm was noted; it was enmeshed in a tenacious fibrous capsule which adhered to the surrounding soft tissues. After the excision of the mass, erosion of the underlying distal and middle phalanx of the fifth toe was noted. On sectioning, the mass showed a yellowish pasty calcareous material and there was a gritty feeling to the material. Upon histological examination, the capsule consisted of a well-defined collagen fiber membrane with an amorphous acidophilic material inside. The mass also contained a crystalline infiltrate of calcium and giant cells. It was identified as an inactive phase of tumoral calcinosis because there was merely calcified material surrounded by dense fibrous material extending into the adjacent tissues [Figure 3]. The patient has been followed up for one year



Figure 2: On enhanced coronal T1-weighted image, this mass reveals multiple nodular areas of low signal intensity suggesting calcifications and peripheral enhancement

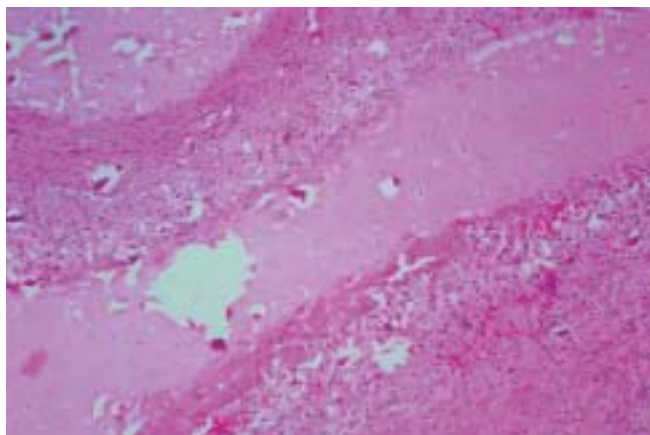


Figure 3: The lesion is an inactive phase of tumoral calcinosis. (H and E, x40)

postoperatively without any evidence of relapse and noted persisting growth of the bone.

## Discussion

Tumoral calcinosis is an uncommon form of extraosseous calcification characterized by large, rubbery or cystic masses occurring mainly in relation to large joints. Prior studies have suggested that tumoral calcinosis most commonly occurs around the hip, shoulder and elbow while it is less frequently seen in the foot and ankle.<sup>[3]</sup>

The exact cause is not known but Smack *et al.*<sup>[4]</sup> formulated a pathogenesis-based classification of tumoral calcinosis into three types. 1. Primary normophosphatemic tumoral calcinosis; in this type patients have no known disorders of phosphate or calcium metabolism. 2. Primary hyperphosphatemic tumoral calcinosis; in this type patients have elevated serum phosphorus and normal serum calcium. The etiology is thought to be a defect in phosphate resorption. 3. Secondary tumoral calcinosis; these patients have a concurrent disease capable of causing soft tissue calcification. These include chronic renal failure with a secondary hyperparathyroidism, hypervitaminosis D, Milk-alkali syndrome and bone destruction.<sup>[1,5]</sup> A high recurrence rate after excision is common in this type. In our patient, no past history or family history of the disease was noted and also serum calcium and phosphorus were within the normal limits and there was no relapse.

Tumoral calcinosis also appears to be triggered by minor trauma. Bleeding is followed by histiocyte aggregation with subsequent formation of cystic cavities lined by the histiocytes.<sup>[3]</sup> The principal manifestation of the disease is the presence of a small-sized firm mass as was in our case. The underlying joints are unaffected and as a rule, the patients are in good health.<sup>[6]</sup>

The calcified juxta-articular mass is the radiological hallmark of tumoral calcinosis and radiological abnormalities are essentially limited to the soft tissues with only the occasional pressure erosion of underlying bony surfaces.<sup>[6]</sup> The adjacent joint space is always preserved. The differential diagnosis of soft-tissue calcification is wide-ranging and must be made with conditions producing soft tissue calcification and ossification, such as osteosarcoma, chondrosarcoma, myositis ossificans or heterotopic bone. With the use of radiography these diseases can be distinguished from tumoral calcinosis in the majority of patients. A biopsy may be warranted in some cases, particularly if musculoskeletal tumors are considered in the differential diagnosis.<sup>[1]</sup> In our case, a radiograph showed significant bony erosions of the distal and middle phalanges of the fifth DIP joint. The patient had undergone excision biopsy and the pathological report identified an inactive phase of tumoral calcinosis of the foot.

Traditionally, treatment was limited to excision, although recurrence was common.<sup>[3]</sup> In our patient, we have been following up for one year without any evidence of relapse. In addition, if there is pressure of calcinosis for an extended period of time, especially for younger patients, it could induce an

underlying bone erosion or damage like in our patient; therefore, active treatment is indicated.

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