

Tumoral calcinosis in the extensor indicis proprius tendon: A case report

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Introduction

Tumoral calcinosis (TC) is a rare clinicopathological entity characterized by the presence of calcified deposits in periarticular soft tissue. The calcified deposits predominantly comprise of calcium hydroxy apatite crystals and amorphous calcium phosphate¹. Clinically, they appear as a painless slowly growing mass in the vicinity of large joints such as hips, shoulder, and elbow^{2, 3}. However, it has also been reported in other areas such as the neck, arms, hands, spine, legs, and feet³. The progressive growth of the mass may result in pain, nerve compression, and functional impairment⁴. The overlying skin is usually intact; however, long-standing lesions may lead to skin ulceration and sinus tract formation with chalk white drainage².

The diagnosis of TC is based on clinical and imaging findings. It has a characteristic radiographic appearance of multilobulated calcification in periarticular soft tissues, commonly on the extensor side of the articulation. Radiolucent lines separate the lobules (fibrous septa)^{5, 6}. Ultrasound can be used to examine lesions, especially with minimal calcification, appearing as heterogeneous multiloculated mass with multiple cavities separated by hyperechoic thin septa. Color doppler may detect blood flow in some of these septa. MRI is superior to CT scan in detecting the extent of the lesions and their relationship with surrounding structures⁶. However, histopathological examination is required for definitive diagnosis. Treatment options include both medical and surgical intervention^{5, 7}.

We present a case of tumoral calcinosis with peculiar presentation. A case of 4-year-old female with tumoral calcinosis of the extensor indicis proprius tendon on the right wrist.

Case

A 4-year-old female presented to plastic and reconstructive surgery clinic with a slowly enlarging mass on the dorsal aspect of the right wrist. The mass was first noted by her parents 2 months ago. It was associated with pain and discomfort, however, there was no functional limitation. Parents denied history of trauma, lesions in other areas of the body, as well as family or medical history of note.

On hand examination, a solitary mass appeared on the dorsal aspect of the right wrist, medial to the lister's tubercle, measuring 1.5 x 1.5 cm. It was a firm, tender mass with intact overlying skin. Range of motion of the wrist was within normal limit. The mass appeared to move with the extension of the right index finger. Sensation was intact and pulses were palpable.

Ultrasound examination revealed a swelling on the dorsal aspect of the wrist measuring 15 x 10 x 5 mm. The swelling was well-defined, multilobulated and mildly echogenic with some vascularity. The swelling was lying within the fourth extensor tendon compartment at the radial aspect and is closely opposed to the extensor indicis proprius tendon^{Figure 1}. Therefore, the working diagnosis was a giant cell tumor of the tendon sheath. No further imaging was performed.

In the operating theatre, exploration of the wrist revealed a mass attached to the extensor indicis proprius tendon. The mass was excised completely and sent for histopathological review. Grossly, the mass consisted of greyish yellow soft tissue, measuring 1.5 x 1 x 0.7 cm. Microscopically, Sections show circumscribed variably sized multinodules separated by fibrous speta; forming lobules contain amorphous granular calcific material with a few scattered psammomatous bodies, and rimmed by histocytes including multinucleated histocytes giant cells. There is no evidence of crystals noted on regular and polarised light microscopic examination ^{Figure 2, 3}. The histopathological report was compatible with tumoral calcinosis.

Consequently, the necessary laboratory investigations were ordered. The results of serum phosphate, serum calcium, vitamin D, parathyroid hormone, renal function test, uric acid and c-reactive protein were within normal limits. Anti-nuclear antibodies were not detected. Thus, a diagnosis of idiopathic tumoral calcinosis was made.

Discussion

Tumoral calcinosis is a rare clinicopathological entity which commonly occurs in the periarticular soft tissue of large joints such as hips (31%), followed by buttock (26%) and upper extremities (15%). In the upper extremities, TC frequently involves the shoulder and elbow, however, the involvement of hand and wrist remain scarce^{3, 8, 9, 10, 11, 12, 13}. Our case presents TC of the extensor indicis proprius tendon. To our knowledge, this is the first case to report TC in the extensor tendon.

Common differential diagnosis of wrist swellings is ganglion cyst, giant cell tumor of tendon sheath and extraskeletal chondroma. Other differentials which may mimic TC, includes tenosynovitis with psammomatous calcifications. Ganglion cyst is a benign condition characterized by mucin filled synovial cyst. It is commonly found on the dorsal aspect of the wrist (70%) arising from scapholunate ligament or articulation. Clinically, both TC and ganglion cyst present as firm painless mass ¹⁴. However, they can be differentiated by ultrasound imaging. On ultrasound, TC appears as heterogeneous multiloculated mass with hyperechoic thin septa. Color doppler may detect blood flow in some of these septa ⁶. On the other hand, ganglion cyst appears as hypoechoic or anechoic well-defined mass with possible septations and acoustic enhancement. No blood flow is detected on color doppler ¹⁵.

Giant cell tumor of the tendon sheath (GCTTS) is the second most common soft tissue tumor of the hand. It originates from the synovium of the tendon sheath and is characterized by a slowly growing, usually painless mass ^{16, 17}. Ultrasound can be used as the first method in diagnosing giant cell tumor as it provides valuable information about the tumor's size, vascularity, and its relation to adjacent structures. GCTTS appears as a solid homogenous hypoechoic mass with detectable vascularity. However, heterogeneity may be seen in minority of the cases ¹⁸. On the contrary, on ultrasound, TC appears as heterogeneous multiloculated mass with multiple cavities limited by echogenic thin septa ⁶. In this case, the ultrasound of the mass revealed a multilobulated and mildly echogenic mass with some vascularity corresponding to GCTTS. Therefore, no further imaging was performed.

Extraskeletal chondroma is benign soft tissue tumor which predominantly occurs in the hands and feet. It presents as a slowly growing mass which results in pain and tenderness. Most are solitary and may be attached to the tendon or tendon sheath. Histologically, it exhibits mature hyaline cartilage arranged in lobules. However, some display calcifications, which mimics TC. Nevertheless, soft tissue chondroma can be distinguished histologically from TC by the presence of cartilage^{8, 19}.

Tenosynovitis with psammomatous calcifications (TCP) is a distinctive variant of idiopathic calcifying tenosynovitis or calcifying tendinitis²⁰. Clinically, TCP presents as a painful mass with predilection to the distal extremities. It usually occurs because of repetitive activity or trauma and is not associated with metabolic abnormalities or family history. Clinically, TC may mimic TCP, however, these two entities are distinguished by histopathological examination. On histopathological examination, TCP is characterized by multiple psammomatous calcifications surrounded by granulomatous infiltrate²¹. In this case the patient presented with atraumatic painful mass on the wrist with no family history or metabolic abnormalities. All together, these clinical features points toward TCP. However, histopathological examination revealed lobules

of calcific material surrounded by histiocytic giant cells with few psammomatous calcifications. Therefore, the final diagnosis was TC.

Clinically, there are three forms of TC: idiopathic, hereditary, and secondary form. The idiopathic form is characterized by onset during the first and second decade of life with a solitary, slowly growing, painless mass. The mass is firmly attached to the underlying structure such as fascia, muscle, or tendon. The familial form is inherited in autosomal recessive manner and characterized by multifocal lesions. It has two variants, hyperphosphatemic and normophosphatemic. The hyperphosphatemic form is distinguished from the normophosphatemic form by elevated levels of phosphate and vitamin D. The secondary form is characterized by the presence of diseases which promote soft tissue calcifications such as chronic renal failure, systemic sclerosis, sarcoidosis, and primary hyperparathyroidism⁸. Our patient presented with solitary calcification with no evidence of familial inheritance. Furthermore, the blood investigations were normal. Based on the preliminary workup, we indicate the absence of underlying metabolic, endocrine and connective tissue disease. Therefore, our case corresponds to idiopathic form of tumoral calcinosis.

Slavin et al. described the histological development of TC based on three stages. Stage one lesion consists of clusters of foamy histocytes aggregates around blood vessels. These histocytes which are found in tendons and fascia, spread into the nearby tendon, fascia, muscle and skin. These histocytes are transformed into cystic cavities as a result of granular necrosis and degenerative alteration in collagen. Stage two consists of cytoplasmic calcification of the disintegrated histocytes and plate-like calcification of the admixed giant cells. Stage three consists of acellular fibrotic tissue filled with dense basophilic calcification⁷. This classification has demonstrated clinical, therapeutic, and prognostic values. Stage one is the active stage and refers to early lesion that only occurs in children. At this stage, the growth is prominent because of proliferative and mitotic activities, resulting in a locally aggressive tumor. Therefore, surgical excision is the mainstay of treatment. On the other hand, stage three, which commonly occur in adults, is unactive. Therefore, it does not require surgical excision unless it cause debilitating symptoms³.

Surgical excision is the mainstay of treatment; however, recurrence is possible⁷. Medical therapy with phosphate lowering agents and calcium and phosphate restricted diet may be helpful^{5, 22}. Our patient was treated with complete surgical excision of the mass.

Conclusion

We conclude that tumoral calcinosis should be considered by surgeons when presented with a wrist swelling. Proper clinical evaluation of the patient aids in the diagnosis and preoperative planning of surgical treatment. Complete surgical excision is the mainstay of treatment in idiopathic form of tumoral calcinosis with low chances of recurrence.

Declaration of consent

Written consent was obtained from the patient's guardian. In the written form, the guardian consent to images and other clinical information used in this report and published in a medical journal. The guardian understands that the patient's name and initials will not be disclosed and that while every effort will be made to conceal her identity, anonymity cannot be guaranteed.

Declaration of conflict of interest

The authors declare that there is no conflict of interest.

Author Contributions

Dr. Maithaa AlShamsi: Writing the manuscript.

Dr. Ammar AlDhamin: Writing and editing the manuscript.

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