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CASE REPORT

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

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Key Clinical Message

Tumoral calcinosis is a rare clinicopathological entity characterized by the presence of calcified deposits in the periarticular soft tissue. Commonly affecting the hips, buttock, shoulders, and elbow, and less commonly in hands, wrists, and feet. We present a novel case of tumoral calcinosis in a 4-year-old female with an atraumatic wrist swelling for 2 months. It was a diagnostic enigma for the surgeon due to the peculiar site of presentation. However, with the help of a pathologist, we diagnosed and successfully treated tumoral calcinosis of the extensor indicis proprius tendon.

KEYWORDS

extensor indicis proprius tendon, extensor tendon, idiopathic, tumoral calcinosis, wrist

1 | 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 phosphate.[1] 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 feet.[3] The progressive growth of the mass may result in pain, nerve compression, and functional impairment.[4] The overlying skin is usually intact; however, long-standing lesions may lead to skin ulceration and sinus tract formation with chalk white drainage.[2]

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 structures.[6] 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.

2 | 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 × 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 1.5 × 1.0 × 0.5 cm. 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 theater, exploration of the wrist revealed a mass attached to the extensor indicis proprius tendon. The mass was excised completely and sent for histopathological review Figure 2. Grossly, the mass consisted of grayish yellow soft tissue, measuring 1.5 × 1 × 0.7 cm. Microscopically, Sections show circumscribed variably sized multinodules separated by fibrous speta; forming lobules. The lobules contain amorphous granular calcific material with a few scattered psammomatous bodies, and rimmed by histocytes including multinucleated histocytes giant cells Figures 3,4. There is no evidence of crystals noted on regular and polarized light microscopic examination. 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. Antinuclear antibodies were not detected. Thus, a diagnosis of idiopathic tumoral calcinosis was made.

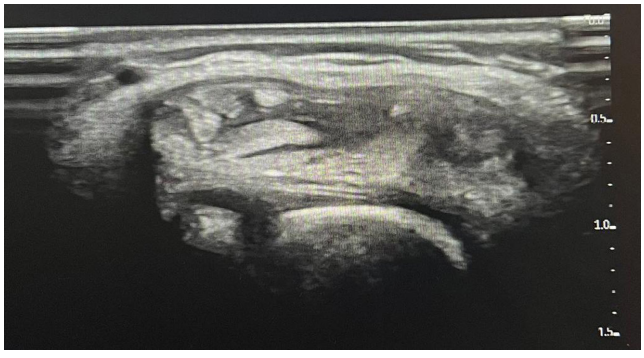

FIGURE 1 Mass attached to the extensor indicis proprius tendon.

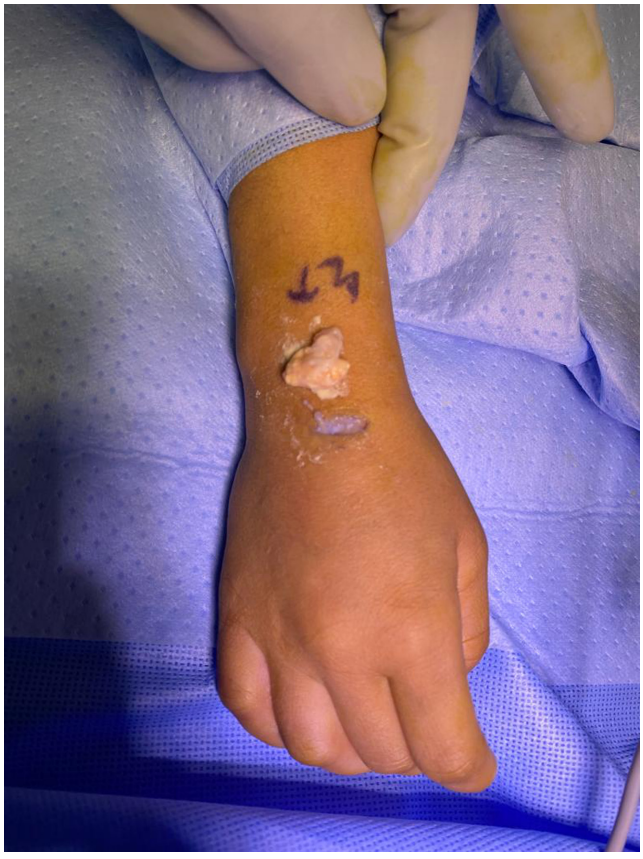

FIGURE 2 Complete excision of the lesion.

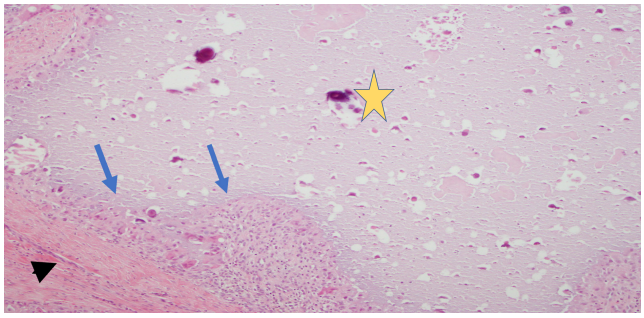

FIGURE 3 Calcified area (yellow star), rimmed by histocytes including multinucleated giant cell histocytes (double blue arrows) and surrounded by fibrosis (black arrowhead).

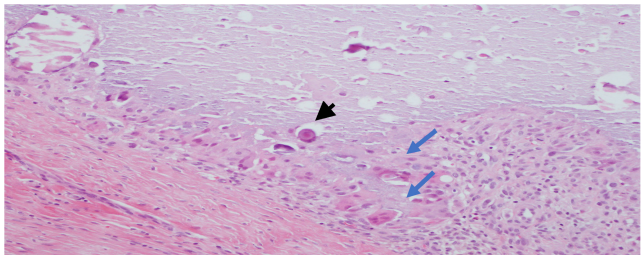

FIGURE 4 Higher magnification of the lesion showing multinucleated histiocytes (blue arrows) and scattered psammomatous calcification (black arrowhead).

3 | 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.[3,8–16] To our knowledge, most of TC hand cases were reported in middle aged patients[9–12,16] However, only few TC hand lesions were reported in children. One case reported in a 7-month-old infant, three cases in children aged 3-, 5-, and 8-year-old and one case in a 15-year-old teenager.[13–16]

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.[17] 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 contrary, ganglion cyst appears as hypoechoic or anechoic well-defined mass with possible septations and acoustic enhancement. No blood flow is detected on color doppler.[18]

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.[19,20] 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.[21] 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.[8,22]

Tenosynovitis with psammomatous calcifications (TCP) is a distinctive variant of idiopathic calcifying tenosynovitis or calcifying tendinitis.[23] Clinically, TPC 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 infiltrate.[24] 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 hyperparathyroidism.[8] 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 calcification.[7] 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 contrary, stage three, which commonly occur in adults, is inactive. Therefore, it does not require surgical excision unless it cause debilitating symptoms.[3]

Surgical excision is the mainstay of treatment; however, recurrence is possible.[7] Medical therapy with phosphate lowering agents and calcium and phosphate restricted diet may be helpful.[5,25] Our patient was treated with complete surgical excision of the mass.

4 | 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.

AUTHOR CONTRIBUTIONS

Maithaa AlShamsi: Writing – original draft. **Ammar AlDhamin:** Writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

The data supporting the findings in this case report is available within the article and its supplementary materials.

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.

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REFERENCES

1. Prasad J, Malua S, Sinha DK, Hassan F, Tekriwal R. Tumoral calcinosis: a case report. *Indian J Surg*. 2007;69(6):251-253. doi:10.1007/s12262-007-0036-y
2. McClatchie S, Bremner AD. Tumoral calcinosis—an unrecognized disease. *Br Med J*. 1969;1(5637):153-155. doi:10.1136/bmj.1.5637.142-a
3. Pakasa NM, Kalengayi RM. Tumoral calcinosis: a clinicopathological study of 111 cases with emphasis on the earliest changes. *Histopathology*. 1997;31(1):18-24. doi:10.1046/j.1365-2559.1997.6050831.x
4. Riemenschneider PA, Ecker A. Sciatica caused by tumoral calcinosis; a case report. *J Neurosurg*. 1952;9(3):304-307. doi:10.3171/jns.1952.9.3.0304
5. Olsen KM, Chew FS. Tumoral calcinosis: pearls, polemics, and alternative possibilities. *Radiographics*. 2006;26(3):871-885. doi:10.1148/rg.263055099
6. Chaabane S, Chelli-Bouaziz M, Jelassi H, Mrad K, Smida M, Ladeb MF. Idiopathic tumoral calcinosis. *Acta Orthop Belg*. 2008;74(6):837-845.
7. Slavin RE, Wen J, Barmada A. Tumoral calcinosis—a pathogenetic overview: a histological and ultrastructural study with a report of two new cases, one in infancy. *Int J Surg Pathol*. 2012;20(5):462-473. doi:10.1177/1066896912444925
8. Goldblum JR, Folpe AL, Weiss SW. *Enzinger and Weiss's Soft Tissue Tumors*. 7th ed. Elsevier; 2020.
9. Hammert WC, Lindsay LR. Tumoral calcinosis—or is it? A case report and review. *Hand*. 2009;4(2):119-122. doi:10.1007/s11552-008-9132-0
10. Kim HS, Suh JS, Kim YH, Park SH. Tumoral calcinosis of the hand: three unusual cases with painful swelling of small joints. *Arch Path Lab*. 2006;130(4):548-551. doi:10.5858/2006-130-548-TCOTHT
11. Gonzalez M, Rettig M, Ayalon O. Extensive Tumoral calcinosis of the hand. *J Hand Surg Am*. 2021;46(11):1031.e1-1031.e6. doi:10.1016/j.jhsa.2020.10.030
12. Sebesta A, Kamineni S, Dumont CE. Idiopathic tumoral calcinosis of the index finger. Case report. *Scand J Plast Reconstr Surg Hand Surg Suppl*. 2000;34(4):405-408. doi:10.1080/028443100750059237
13. Evans DM, Lewis JS. Tumoral calcinosis associated with congenital malformations of the hand. *J Hand Surg Br*. 1994;19(5):647-652. doi:10.1016/0266-7681(94)90136-8
14. Murai S, Matsui M, Nakamura A. Tumoral calcinosis in both index fingers: a case report. *Scand J Plast Reconstr Surg Hand Surg Suppl*. 2001;35(4):433-435. doi:10.1080/028443101317149426
15. Amadei F, Petrolati M. Idiopathic tumoral calcinosis associated with congenital malformation of the hand. Case report. *Ann Chir Main Memb Super*. 1998;17(1):59-62. doi:10.1016/s0753-9053(98)80021-5

16. Malik M, Acharya S. Tumoral calcinosis of the fingers: a report of two cases. *Int Orthop*. 1993;17:279-281.
17. Gregush RE, Habusta SF. *Ganglion cyst*. StatPearls; 2022. https://www.ncbi.nlm.nih.gov/books/NBK470168/
18. Wang G, Jacobson JA, Feng FY, Girish G, Caoili EM, Brandon C. Sonography of wrist ganglion cysts. *J Ultrasound Med*. 2007;26:1323-1328. doi:10.7863/jum.2007.26.10.1323
19. Hasham S, Burke FD. Diagnosis and treatment of swellings in the hand. *Postgrad Med J*. 2007;83(979):296-300. doi:10.1136/pgmj.2005.043992
20. Darwish FM, Haddad WH. Giant cell tumour of tendon sheath: experience with 52 cases. *Singapore Med J*. 2008;49(11):879-882.
21. Middleton WD, Patel V, Teefey SA, Boyer MI. Giant cell tumors of the tendon sheath: analysis of sonographic findings. *AJR Am J Roentgenol*. 2004;183(2):337-339. doi:10.2214/ajr.183.2.1830337
22. Le Corroller T, Bouvier-Labit C, Champsaur P. Diffuse mineralization of forearm extraskeletal chondroma. *Joint Bone Spine*. 2008;75(4):479-481. doi:10.1016/j.jbspin.2007.06.019
23. Gravanis MB, Gaffney EF. Idiopathic calcifying tenosynovitis. Histopathologic features and possible pathogenesis. *Am J Surg Pathol*. 1983;7(4):357-361.
24. Michal M, Agaimy A, Folpe AL, et al. Tenosynovitis with Psammomatous calcifications. *Am J Surg Pathol*. 2019;43(2):261-267. doi:10.1097/pas.0000000000001182
25. Varma S, Bhansali A, Gill SS, Kumari S, Singh S. Peri-articular 'hard tumours' in soft tissues. *Postgrad Med J*. 1999;75(886):492-495. doi:10.1136/pgmj.75.886.492

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