Tumoral Calcinosis Of Hand: A Rare Location With Unusual Presentation

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Citation

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Abstract

Tumoral Calcinosis is an entity characterised by large peri-articualar deposition of calcium phosphate that resembles a neoplasm. Although the aetiology remains obscure, this clinical entity is well established in literature. More than 300 case reports have been published in English literature, but isolated involvement of hand is extremely rare. We herein report a rare case of Tumoral Calcinosis in hand with a very unusual presentation. The clinical presentation posed a diagnostic problem, as it appeared to be infection till the radiograph revealed calcified nodular deposits. The calcific deposits were successfully removed surgically and patient had dramatic relief of symptoms. There was no disturbance of Calcium or phosphate metabolism and no family history of similar disorder was present. After two years of follow up, there was no recurrence and patient is symptom free.

INTRODUCTION

Any abnormal deposition of calcium salt in the soft tissue is called Calcinosis [1,2]. There are three varieties of calcinosis [1,2], viz.:

1. Calcinosis universalis
2. Calcinosis circumscripta
3. Tumoral calcinosis

Calcinosis universalis is confined to children with deposition of calcium salts as nodules or plaques in skin, subcutaneous tissues and superficial muscles. This is associated with scleroderma in 40% of cases. The prognosis is usually bad.

Calcinosis circumscripta has a benign course and occurs in middle-aged women. The deposits are small, nodular or streaky and affect the flexor tendon sheaths of hands and wrist. Scleroderma, sclerodactyly or Raynaud's disease is associated with 30-40% of cases.

The third variety is rare and is characterised by large nodular peri-articualar deposits of calcium phosphate that resembles a neoplasm. Inclan (1943) [3] named it as “Tumoral calcinosis”. This lesion is different from the universal and circumscribed types. The lesions are more common among blacks and about 2/3rd of the cases reported are in blacks. More than 1/2 of the patients have an affected sibling. It is usually seen in adolescents and young adults. The lesions are usually asymptomatic and only rarely cause discomfort, pain and tenderness. The underlying joints are unaffected and as a rule the patients are in a good general health. This usually results in delayed presentation when the lesions have attained a large size. The masses usually are firmly attached to the underlying fascia, muscle or tendon. Rarely the lesion might ulcerate and discharge yellow-white chalky fluid, it may get secondarily infected and result in a fistula.

There has been a lot of confusion about the aetiology of this condition, which has resulted in many names for the same condition. Although the aetiology is not yet known, the term Tumoral calcinosis is generally accepted to describe this condition. It is classically known to affect large joints and involvement of hand is extremely rare. We herein report such a rare case of Tumoral calcinosis in hand. The presentation of this case was like an infection, which makes this case a unique one. The aim of this report is to make the hand surgeons aware about the unusual presentation of this rare condition, which can cause a diagnostic confusion.

CASE REPORT

A 22 years old housewife, who was apparently healthy, presented with complaints of severe pain and swelling over the second web space of seven days duration. She noticed swelling seven days back associated with pain, which worsened rapidly in next five days. There were no other such lesions in the body. Patient reported to a local general

physician who suspected infection and referred the patient to us. The pain was severe enough to prevent her using the hand. But surprisingly, the patient was afebrile and in good general health. At first instance it appeared to be infection but the absence of fever, and general good health raised suspicion in our mind. There were no signs of tenosynovitis, palmer space infection or flexor sheath infection. There was no regional lymphadenopathy. Movements at the metacarpo-phalangeal joint were possible, but range was decreased. Movements within the range possible were painless.

There was no history of similar complaint in the past. There was no family history of similar problem.

Radiograph of the hand showed multiple round to oval, well-demarcated masses of calcification, located in the peri-articular soft tissue of the second metacarpo-phalangeal joint. The calcified masses appeared lobulated and seemed to consist of a conglomeration of multiple small and round opacities, unattached to bone. The joint was essentially normal with smooth articular margin and good joint space.

All the blood investigations including the ESR & cell counts were within the normal range and there was no evidence of infection in the blood tests. Calcium, phosphorus & alkaline phosphatase levels also were normal.

Surgical excision of the calcific deposits was done though the dorsal approach to the second metacarpo-phalangeal joint. The calcific masses were hard and attached to the deeper tissues; they lay close to the tendon sheath of the flexor tendons and at places were infiltrating it. On sectioning, the mass showed yellowish pasty calcareous material and there was a gritty sensation. There was no involvement of the metacarpo-phalangeal joint. We could excise the whole calcified mass with tedious dissection.

Till two years of regular follow up, there was no evidence of any recurrence or similar lesions in any other part of the body.

Figure 1

Figure 1: Preoperative photographs of the patient showing the swelling in the region of the second web space.

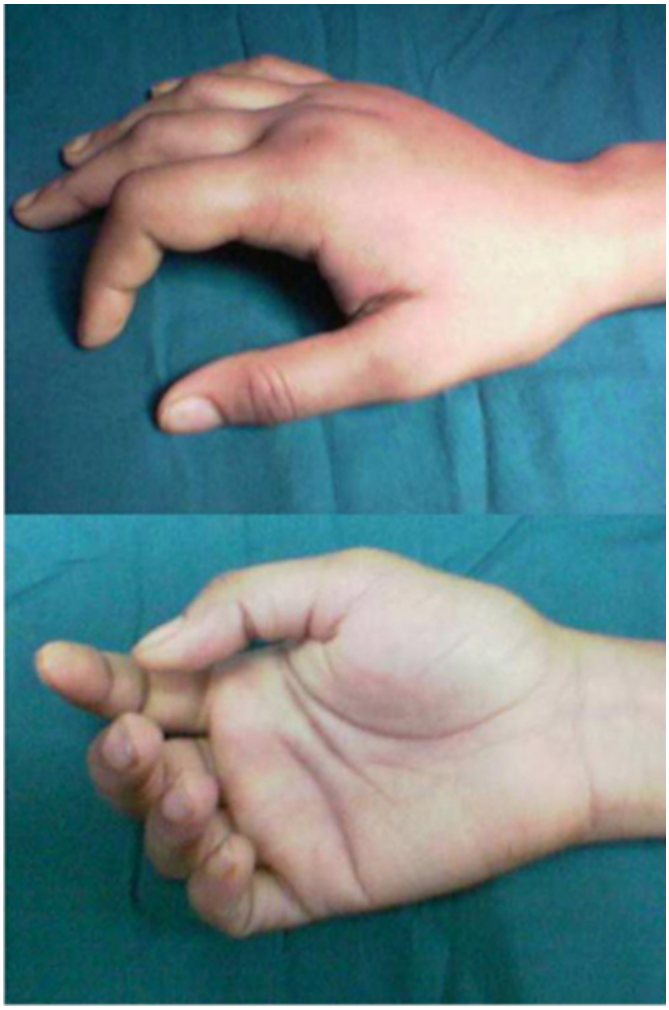


Figure 2

Figure 2: Anteroposterior and lateral radiograph of the hand showing the round to oval lobulated masses of calcification in the second web space.

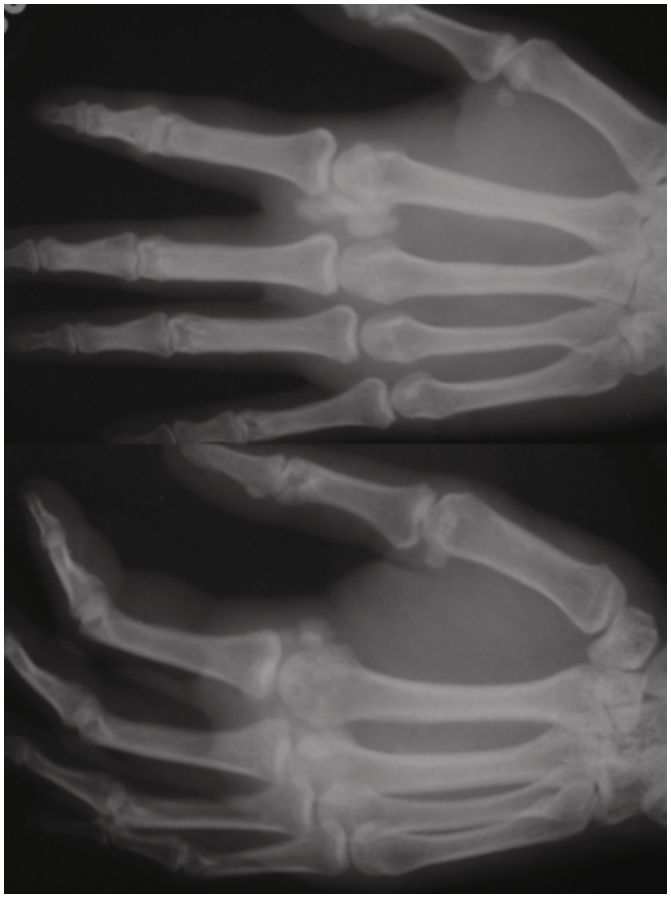
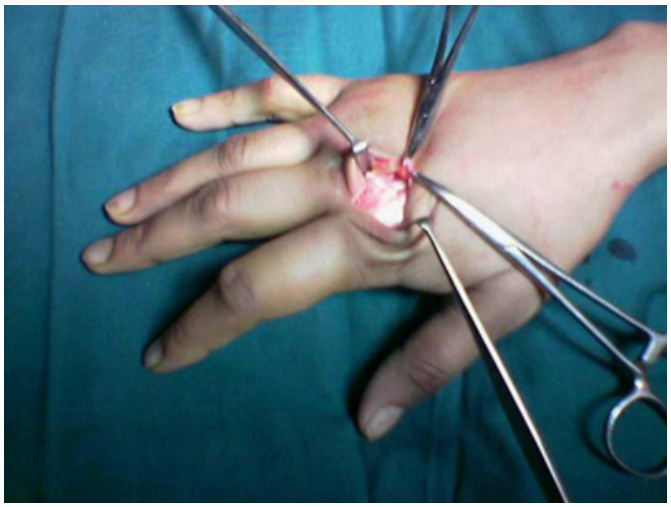


Figure 3

Figure 3: Intraoperative photograph of the patient showing the calcified masses.



DISCUSSION

First report of Tumoral calcinosis was by Duret in 1899 [4]. He described a 17 years old girl who had calcific tumor of the elbow and buttocks and called it as “endotheliome calcifie”. Teutschlander (1935) described a case and thought that the calcification followed fat necrosis and called the lesion “calcium tumor” due to lipocalcinogranulomatosis [4]. Other terminologies, which have been assigned to this obscure condition, are: Calcifying bursitis; Calcifying Collagenolysis; Kikuyu bursa [5], but the most widely accepted terminology is ‘Tumoral Calcinosis'. It was Inclav in 1943, who first introduced the term “Tumoral Calcinosis” [3], to describe the deposition of prominent calcified masses overlying large joints, such as the hips, elbow and shoulders. He reported 3 cases in Negro boys. Two theories have been put forward for its occurrence [5] -inborn error of phosphorus metabolism & mechanical trauma or repeated minor injuries. The disease is thought to be a result of metaplasia of connective tissue cells and not of disturbed calcium or phosphorus metabolism [6]. The disease has a genetic background and the primary defect is in the collagen, which undergoes calcification [5].

Tumoral calcinosis occurs in younger patients, affects multiple joints and has a familial tendency. Sites of preference [1] are lateral and superior aspect of shoulder, posterior aspect of elbow and laterally about the hips and buttocks. Less common sites are feet, retroscapular, acromioclavicular sacral and ischial region. It may occasional involve neck. Very rare sites are hand and distal femur. There are plenty of Tumoral calcinosis cases reported in the literature but hand involvement is only rarely reported, moreover isolated hand involvement is extremely rare. Barton [1]reported three cases of tumoral calcinosis but none of the cases had involvement of the hand. Laffert[7] reported 18 patients of tumoral calcinosis. One patient had involvement of the wrist but none had hand involvement. Smit and Schmaman[8] reported two cases with extensive involvement but none had hands involved. Harkess[4] reviewed the literature before 1967 and reported total of 33 cases including six of his own. Only one patient out of 33 had involvement of the hand. Baldursson[6] reported four cases in a single family but hand involvement was absent in all. Hacihanefioglu[9] reported 11 definite and 9 probable cases of tumoral calcinosis. Only one patient out of all had involvement of finger. Malik & Acharya[10] (1993) reported two cases of tumoral calcinosis involving the fingers. Sabesta, Kamineni & Dumont[11] in 2000 reported a case of idiopathic tumoral calcinosis of the index finger. Murali, Matsui & Nakamura[12](2001) reported an interesting case of

tumoral calcinosis with bilateral involvement of index fingers in a five-year-old child. Kuhlmann, Rambani, Dhillon & Aggarwal [5] reported a case of multiple para-articular calcific masses involving the elbow, hand, back, knee and the feet. They stressed that the distal location of the swellings in their case was an unusual feature. Mimoun & Baux[13] (Article in French) in 2004 reported a case of erosive type of tumoral calcinosis affecting the long finger.

Tumoral calcinosis classically has a very benign clinical course. It presents as asymptomatic and slowly growing masses. Its acute presentation, which mimics infection, is very atypical. Sabesta, Kamineni & Durmont[11] reported a case of finger involvement, which appeared like infection and posed a diagnostic problem. Our case also presented as infection, but the general good health and normal blood investigations prompted us to look for other cause. All the serological investigations were normal in the presented case.

Differential diagnosis of tumoral calcinosis includes: calcinosis universalis; calcinosis circumscripta; vitamin-D intoxication; Burnett's syndrome; renal osteodystrophy and pseudogout. It can be differentiated from the deposits in Pseudogout by the nature of crystals on X-ray, which are lobular in Tumoral calcinosis but granular and more delicate in Pseudogout. X-ray diffraction analysis is confirmatory.

The calcium deposits are characteristically peri-articualar (1-20cm or more in size). They are dense, irregular and have a round or oval contour. Radiolucent fibrous septa separate clumps of calcium, giving a multinodular appearance. There are no bony abnormalities.

Pathology: Multilocular structure with fibrous connective tissue bands separating pasty, calcareous deposits. On microscopy macrophages, multinucleated giant cells and chronic inflammatory cells lining the fibrous bands are noted.

Complications are rare, but there may be ulceration of the overlying skin with secondary infection, fistula formation and discharge of yellow-white chalky fluid.

Surgical excision is the mainstay of treatment. It should be undertaken early as it is difficult to remove a larger lesion and will necessitate more extensive surgical procedure. Recurrence after surgical excision is a problem, but can be managed by repeat excision.

SUMMARY

A Case of Tumoral Calcinosis affecting metacarpo-phalangeal joint region has been described. This is a very rare site for Tumoral calcinosis, which is classically known to affect larger joints. Most interesting thing about the case presented is its mode of presentation. The onset was very acute and at presentation it appeared to be infection. Such an unusual presentation of tumoral calcinosis at an extremely rare site like can pose a diagnostic confusion. This report is to make hand surgeons aware of this unusual presentation of tumoral calcinosis in hand.

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