

Case Report

Tophaceous pseudogout Occurring in the Big Toe - a Report of Two Cases

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Citation: Tanaka S, Hatori M, Noguchi K, Itoi E (2016) Tophaceous pseudogout occurring in the Big Toe - A Report of Two Cases. Ann Case Rep 2017: G137. DOI: 10.29011/2574-7754/100037

Received Date: 29 December, 2016; **Accepted Date:** 09 January, 2017; **Published Date:** 14 January, 2017

Abstract

Peri articular massive focal Calcium pyrophosphate dehydrate crystal deposition (Tophaceous pseudogout) at the small joints is uncommon. We report radiological and histological features of Tophaceous pseudogout arising in the big toe Metatarsophalangeal (MTP) joint. The patients were 48 and 69-year-old female. Their chief complaint was pain in their big toe. Radiological examination showed calcification juxtaposed to the proximal phalanx. MR imaging revealed an irregular-shaped tumor with low signal intensities on the T1 weighted images and high signal intensities on the T2 weighted images. The peripheral area of the tumor was enhanced after gadolinium agent injection. Histologically it was diagnosed as pseudogout. Symptoms disappeared after tumor removal. We should suspect tophaceous pseudogout as one of the differential diagnoses in case of painful calcification in the toe.

Introduction

Calcium Pyrophosphate Dehydrate (CPPD) crystals were first identified in synovial fluid exudates of patients who had acute gout-like arthritis without sodium urate crystals. This entity was defined as Pseudogout. Various clinical features associated with CPPD mimic those of gout, rheumatoid arthritis and so on [1-3]. Pseudogout is a disease where CPPD crystals become deposited on the articular cartilages and the surrounding tissues causing arthritic symptoms. It mainly occurs in the large joints, and infrequently in the small joints such as the phalanx of the toe. We report two cases of tophaceous pseudogout (Tumoral Calcium Pyrophosphate Crystal Deposition Disease [4] which developed on the proximal phalanx of the big toe.

Case 1

A 48 year old woman complained of severe pain in her left big toe. There was no trauma history. She had a conservative treat-

ment with NSAIDs (Non-Steroidal Anti-Inflammatory Drugs) by her previous doctor as hallux valgus but her symptoms remained unimproved. She was referred to our hospital. Physical examination showed a palpable mass, local tenderness, swelling in the big toe MTP joint (Figure 1). CRP level was 0.56 mg/dl. Uric acid level was normal. Plain X-Ray and CT scan showed calcification in the big toe MTP joint (Figure 2). MR imaging showed an irregular-shaped tumor showing low signal intensities on the T1 weighted images and high signal intensities on the T2 weighted images. The peripheral area of the tumor was enhanced with gadolinium agent injection but there were no clear contrast effects seen inside of the tumor (Figure 3 a,b,c). Longitudinal skin incision on the lateral side of the extensor tendon exposed a lesion. The tumor with white granules, attached to the cortex of the proximal phalanx was curetted (Figure 4). Basophilic crystal deposition with peripheral cartilaginous metaplasia was seen histologically. The cartilaginous cells with atypical nuclei was observed. This lesion was diagnosed as CPPD crystal deposition disease (Figure 5). Symptoms disappeared after tumor removal.



Figure 1: Macroscopic views of the left foot showing mild swelling (arrows).

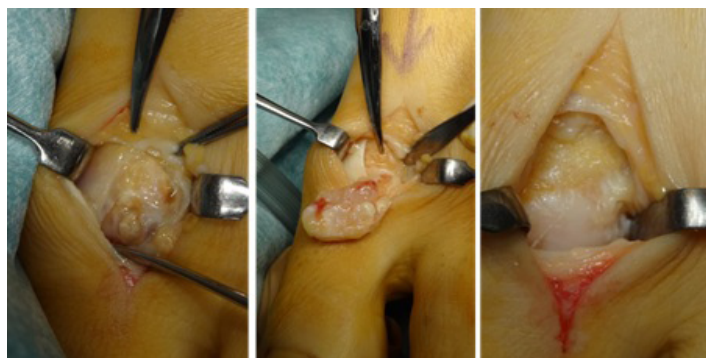


Figure 4: Intraoperative views. The tumor had white granular appearance, attached to the cortex of the proximal phalanx.

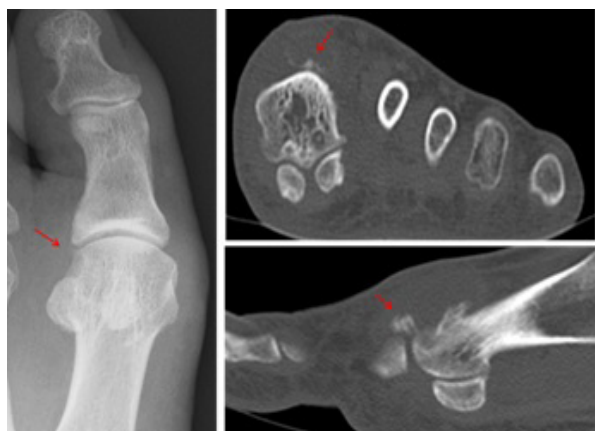


Figure 2: Plain X-Ray and CT scan showing calcification of the proximal phalanx (arrows).

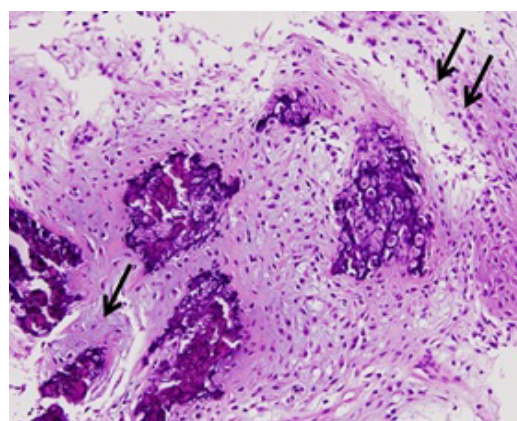


Figure 5: Pathological findings. Deposition of basophilic crystals was observed (arrow). Cartilaginous metaplasia was seen clearly around the crystals. (arrows).

Case 2

A 69-year-old woman suddenly had pain in the right big toe. She had a conservative treatment with NSAIDs by her previous doctor as hallux valgus but her symptoms remained unchanged. She was referred to our hospital.

Physical examination revealed a palpable mass on the lateral side of the right big toe. No local heat was found. There were no abnormalities of blood examination. Plain radiograph showed lateral cortex erosion of the big toe proximal phalanx (Figure 6a). MR imaging showed an irregular-shaped tumor with low signal intensities on the T1 weighted images and high signal intensities on the T2 weighted images. After intravenous gadolinium injection, only peripheral area was enhanced. (Figure 6 b,c,d).

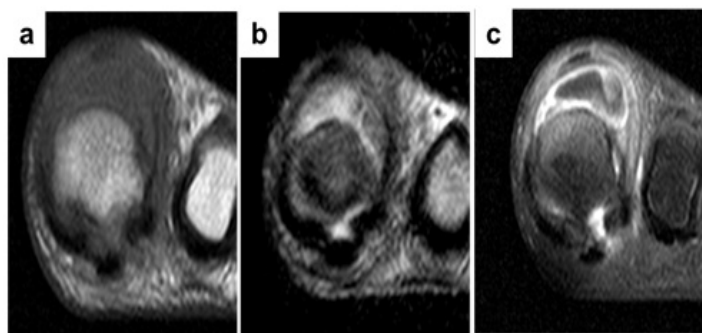


Figure 3: (a) Axial T1 weighted images (b) Axial T2 weighted images. (c) images taken using a gadolinium contrast agent with a fat suppression method.

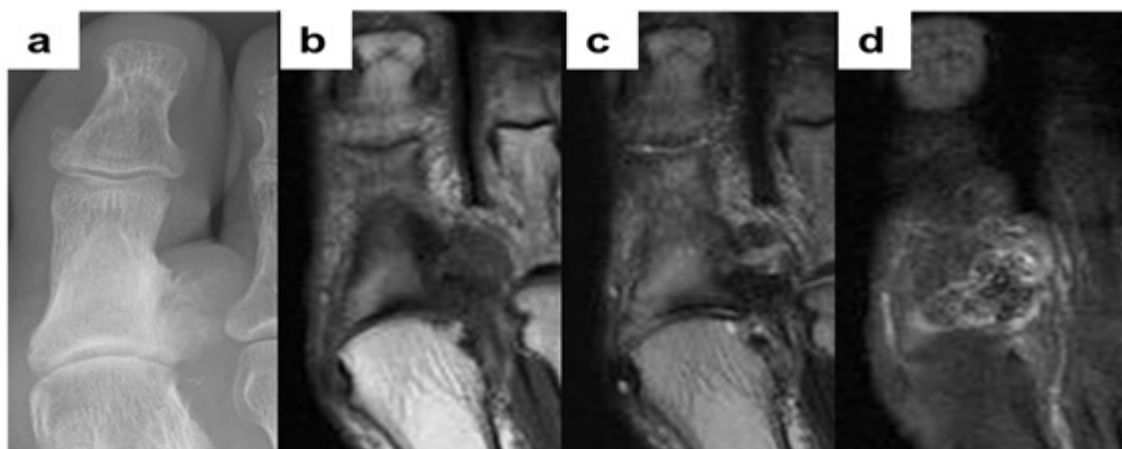


Figure 6: (a) Radiograph of the right big toe. The irregular lateral side of proximal phalanx of big toe and the calcification expanded to the outside of the proximal phalanx is found. (b) showing low signal intensities on the T1 weighted images (c) showing high signal intensities on the T2 weighted images (c) After the gadolinium contrast agent injection the peripheral area was enhanced.

At surgery a tumor with white granules, attached to the proximal phalanx was curetted. The cortex was scalloped and no intramedullary invasion (Figure 7). Histological examination showed basophilic crystal deposition with pavement-like arrangements and granular shapes. Cartilaginous metaplasia was seen clearly around the crystals. Enlarged cartilaginous cells with swollen and atypical nuclei were observed. Based on the crystal shapes and prominent cartilaginous metaplasia, this case was diagnosed as CPPD crystal deposition disease (Figure 8). Symptoms disappeared after tumor removal.

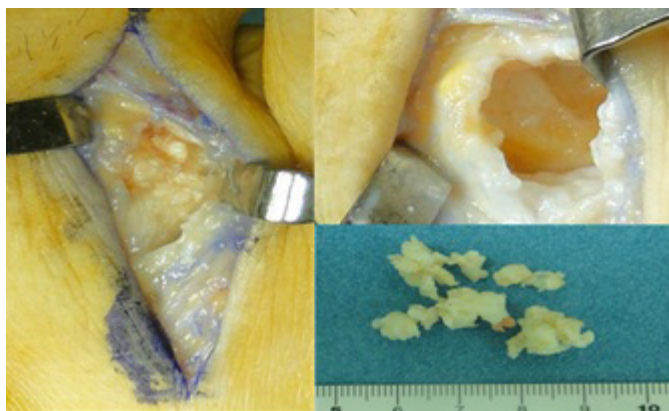


Figure 7: Intraoperative findings. The tumor was white granular appearance, attached to the cortex of proximal phalanx. The cortex was scalloped and no intramedullary invasion.

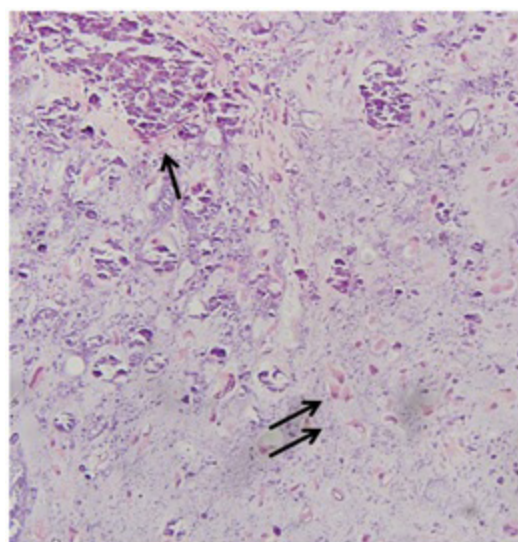


Figure 8: Pathological findings. Deposition of basophilic crystal was observed (arrow). Cartilaginous metaplasia was seen clearly around the crystals (arrows).

Discussion

CPPD deposition disease shows various clinical conditions [3]. The diseases were first identified in synovial fluid exudates of patients with the Pseudogout syndrome; the crystals had induced an acute inflammatory response similar to the urates seen in gout

[1]. It is a relatively common arthritic disorder in the elderly and the majority of the affected joints were the knee, wrist and pubic bone [5]. However tophaceous pseudogout (tumoral or massive CPPD crystal deposition disease) in the articular and para articular is rare [4]. Phot Luisiri reviewed the literatures saying 14 patients with tumoral calcifications consisting of CPPD rather than basic calcium phosphate [6]. The described patients had a lesion in the big toe.

It was reported the clinical features of the patients with tophaceous pseudogout were pain, painful mass, mass, swelling [1]. Although occurrence of CPPD in the foot is very rare, it must be considered one of the differential diagnoses in case of patients especially women presenting inflammatory arthropathies with or without radiographic signs [7]. Serum calcium and uric acid level was normal in tophaceous pseudogout [4-6]. Our reported cases had a painful mass in the toes without other arthritis or metabolic diseases that frequently accompanies CPPD deposition diseases.

The radiographic features of tophaceous pseudogout were described as considerable subcutaneous periarticular calcification on the X-ray, the lesion appeared as a slightly or densely calcified mass; the pattern of calcification was granular or fluffy [4-6]. The present CT scan is useful to localize the mass location and erosion on the bone surface. MR imaging showed low signal intensities on the T1 weighted images and high signal intensities on the T2 weighted images. The peripheral area of the tumors was enhanced, but there were no clear contrast effects seen inside the tumors, which is the feature of this tumor.

The differentiation diagnosis is hallux valgus, goutly attack, septic arthritis and benign or malignant tumors [4,7-9]. Our patients were treated as hallux valgus because of the pain and swelling in their first MTP joint. Blood examination is useful to distinguish gout nodes from the other diseases. It is no worth that uric acid level was normal in tophaceous pseudogout.

Surgery is treatment of choice to heel and confirm the diagnosis, once the acute symptoms abate, if there is no therapeutic method to slow the process of CPPD deposition [7]. In our cases surgical treatment was performed. The mass was curetted and their symptoms was disappeared.

The histological feature was small or large deposition of basophilic calcified materials containing needle shaped and rhomboid crystals, foreign body granulomatous reaction to the CPPD, chondroid metaplasia was observed [4]. In our cases similar features were found, that was cartilaginous metaplasia and enlarged cartilaginous cells with swollen and atypical nuclei was seen clearly around the crystals. Needle aspiration is effective to diagnose preoperatively [6].

In conclusion, when a painful calcification lesion is observed in the toe, pseudogout nodes was suspected as one of the differential diagnoses. Surgical treatments were effective heel and make an accurate diagnosis of the disease.

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