Tumoral calcinosis misdiagnosed

Win Lin Chai[1], Yuen Hoong Phang[1], Hwee Cheng Chong[2]

Abstract

Tumoral calcinosis is an uncommon condition which has been described to exist in primary and secondary forms. A lack of awareness of this entity can lead to unnecessary procedures and incorrect management. We report a case of a patient on peritoneal dialysis who presented with multiple painful joint swellings to the orthopaedic department. An initial diagnosis of septic arthritis was made, then revised to chronic tophaceous gout and referred to the rheumatology unit.

IeJSME 2018 12(1): 18-20

Keywords: Dialysis, gout, orthopaedics, rheumatology, tumoral calcinosis, joint

Introduction

Tumoral calcinosis refers to massive periarticular calcinosis. It is an under-recognised condition in our local setting which has been described to exist in primary and secondary forms; the latter occurring in conditions that promote ectopic calcifications, such as renal failure with secondary hyperparathyroidism, primary hyperparathyroidism, systemic sclerosis among many others.[1]

Due to its rarity, it remains a debate if the terminology of tumoral calcinosis should be restricted to the original description of the hereditary form or broadly used to encompass all periarticular calcifications.[2]

The diagnosis is made via its classical radiological and histopathological appearances, supported by clinical and biochemical findings.[3] This condition has also been reported to have various mimics, therefore making an accurate diagnosis can be challenging.[2]

Case Presentation

A 30-year-old Malay gentleman presented with a five-month history of multiple gradually enlarging swellings which became painful a month prior to his hospital admission. He has type two diabetes mellitus and hypertension for thirteen years and commenced on peritoneal dialysis for end stage renal failure in the recent three years. There was no significant family history of joint disorders.

Examination revealed swellings over both shoulders, elbows, wrists, right first metacarpophalangeal joint and left hip. The largest swelling was over the left shoulder - soft, immobile, tender and measured 10cm by 10cm. The other swellings ranged from 2cm to 5cm in diameter. He was afebrile.

Needle aspiration of the left shoulder and left elbow swellings yielded 100ml and 20ml of creamy white material respectively, presumed to be purulent in nature. He subsequently underwent incision and drainage of these swellings under general anaesthesia. Intraoperative findings noted purulent discharge and chalky white material from both sites; joint capsules were uninvolved.

Reports of intraoperative specimens were subsequently negative for bacterium, mycobacterium and fungus. Histopathological examination noted abundant areas of calcification surrounded by fibrotic tissue infiltrated by lymphocytes, plasma cells and numerous multinucleated giant cells with area of proliferation of blood vessels. No granuloma or malignancy was seen.

This patient was referred postoperatively to the rheumatology unit for further management of gout. On review of the joint radiographs, soft tissue calcifications were noted to be extensive with relatively normal bones and joints, which was inconsistent with a diagnosis of chronic tophaceous gout. Polarised microscopy of aspirate material from a soft tissue swelling on the left wrist did not reveal any uric acid crystals. Radiologist verification was sought and a diagnosis of tumoral calcinosis was given (Figure 1).

A plain computed tomography was later done to further define the radiographic appearance of this condition which was best described in the right shoulder (Figure 2). CT revealed multi-lobulated calcification surrounding the right shoulder joint with no intraarticular extension and no bony erosion. There is a cystic component with some cysts showing fluid-calcium levels, typical of tumoral calcinosis.

[1]Department of Medicine, Hospital Melaka, Melaka, MALAYSIA

[2]Physician and Rheumatologist, Department of Medicine, Hospital Melaka, Melaka, MALAYSIA

Address for Correspondence:

Dr. Win Lin Chai, Department of Medicine, Hospital Melaka, Jalan Mufti Haji Khalil, 75400 Melaka.

Email: cwinlin@gmail.com; Tel no.: 6012-2046290; Fax number: 06-2827501

Laboratory examinations showed normal corrected serum calcium [2.34 mmol/L (normal range 2.18-2.60)], high phosphate [3.14 mmol/L (normal range 0.78-1.65)], mildly elevated intact parathyroid hormone (iPTH) [55 pg/ml (normal range 5-39)] and slightly raised uric acid [600micromol/L (normal range 220-547)].

Discussion

The differential diagnoses of painful joint swellings in a patient with renal failure would commonly be septic arthritis due to an immunocompromised state and exposure to repeated invasive procedures, or gout as a result of impaired renal handling of urate. Our patient did not report any fever, thus septic arthritis was not impossible but less likely. Tophaceous gout was a reasonable consideration.

Tumoral calcinosis is not a common cause of joint swelling and is usually not painful; hence the initial misdiagnosis, which led to incision and drainage of multiple sites in our patient. Although tumoral calcinosis is often a radiological diagnosis, any periarticular swelling in a renal failure patient should raise a clinical suspicion for this condition.

Our patient is likely to have secondary tumoral calcinosis as he has renal failure on peritoneal dialysis. However, his biochemical profile was only mildly deranged and differed from cases reported in the literature where the abnormalities were more pronounced. Genetic testing is not available locally to exclude the primary form.

Different diagnoses require different management strategies. Inaccurate diagnosis may result in unnecessary procedures, delay in treatment and possibility of treatment-related complications. Surgery is not usually recommended in the management of tumoral calcinosis, as these swellings tend to be recurrent[4]. There is occasionally a role for surgery in improving joint mobility, as tumoral calcinosis may present with complications, such as, pain due to nerve compression, ulceration, bacterial infection and cosmetic disfigurement[5]. Similarly, surgery is not advocated for uncomplicated tophi. Management of tophaceous gout in a patient with renal impairment requires a careful introduction and up-titration of urate lowering agents.

Conclusion

Tumoral calcinosis should be included in the differential diagnosis of joint swelling in a patient with advanced renal failure. An improved awareness will prevent unnecessary procedures which may result in further debilitation.

Learning points for clinicians

Tumoral calcinosis is uncommon but a clinician may suspect this diagnosis especially in patients with conditions that predispose to ectopic calcifications. The lack of joint destruction in the presence of extensive soft tissue calcifications on plain radiographs should alert the clinician to this disease entity. Septic arthritis and chronic gout cause joint and bone damage, which are evident on various imaging modalities.

Acknowledgement

We would like to acknowledge the contribution of Dr Chong Aun Kee and Dr Rozalina Abd Hamid towards the diagnosis and their assistance with the enhanced radiological images. We thank the Director General, Ministry of Health for his approval of this manuscript for publication.

REFERENCES

1. Goldblum JR, Weiss SW, Folpe AL. Enzinger and Weiss's soft tissue tumors. 6th Ed. Saunders; 2014: 947.
2. Kathryn M. Olsen, Felix S. Chew. Tumoral calcinosis: pearls, polemics and alternative possibilities. Radio Graphics 2006 May-June; 26 (3). Published online 2006. doi.org/10/1148/rg.263055099
3. Ibrahim Fathi, Mahmoud Sakr. Review of Tumoral Calcinosis: A rare clinico-pathological entity. World Journal Of Clinical Cases Sep 2014; 2(9): 409-14. Published online 2014 Sep 16. doi: 10.12998/wjcc.v2.i9.409;PMCID: PMC4163761
4. Hiren Patel, Hamid Shaaban, Howard Benn. A rare case report of tumoral calcinosis syndrome in an adult with rheumatoid arthritis. Clin Cases Miner Bone Metab. 2015 Jan-Apr; 12(1): 62–4. Published online 2015 Jun 1. doi: 10.11138/ccmbm/2015.12.1.062;PMCID: PMC4469231
5. Saumyaranjan Mallick, Zohra Ahmad, Arun K Gupta, Sandeep R Mathur. Hyperphosphatemic tumoral calcinosis. BMJ Case Rep. 2013; 2013: bcr2013008728.

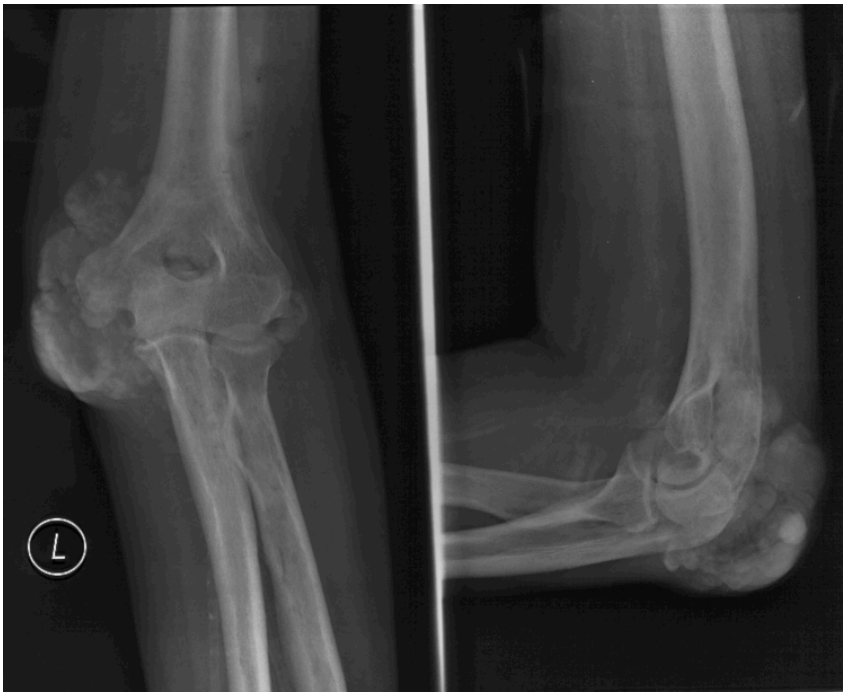

Figure 1: Radiograph of left elbow

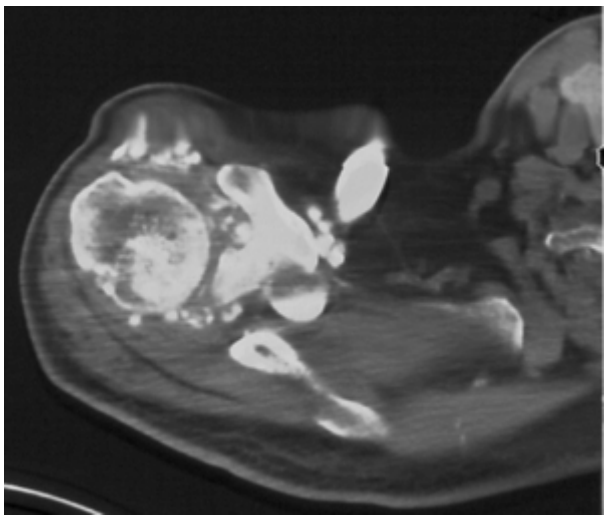

Figure 2: Computed tomography scan of the right shoulder