

Severe tumoral calcinosis in patient with chronic kidney disease

kawtar nassar (✉ kawtamassar@yahoo.fr)

Rheumatology department. Ibn Rochd University Hospital. Hassan II University of Medicine and pharmacy. Casablanca. Morocco <https://orcid.org/0000-0002-1879-2363>

saadia janani

Rheumatology department. Ibn Rochd University Hospital. Hassan II University of Medicine and pharmacy. Casablanca. Morocco

Case Report

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Abstract

Tumoral calcinosis is a rare condition characterized by the deposition of calcium material in extra-articular soft tissues taking a tumoral form. It can be primary or secondary. The chronic kidney disease is among cause of metastatic calcifications but the extensive periarticular cyst-like calcium salt containing space-consuming lesions in the form of tumoral calcinosis is very rare. Different factors predispose the dialysis patient to calcium salt deposits. We report a case of 61 years-old-patient with chronic kidney disease under periodic hemodialysis due to hypertensive nephropathy who developed mechanical pain, evolving since three months, in the left hip, the upper extremity of the right leg and both shoulders with swelling and limited movement interfering with daily activity. The severe TC was diagnosed by the radiography and MRI requiring the modification of treatment strategy.

Introduction

Tumoral calcinosis (TC), a very rare disease entity, occurred in the described patient with end-stage renal disease (ESRD) due to disturbed calcium phosphate metabolism, in which a calcified mass grows around a large joint, and can occur in patients undergoing renal dialysis (1). It is characterized by the deposition of calcium material in the periarticular soft tissues. It can be familial, idiopathic or secondary, especially to chronic kidney disease with or without hemodialysis (2). Clinically, the swelling is firm, mobile in relation to the skin, usually asymptomatic, it can be responsible for joint stiffness and bone pain (3). It can be single or multiple. The diagnosis is radiological, made by plain radiography. Ultrasound shows a polylobed, hyperechoic appearance with a characteristic calcified peripheral shell. Magnetic resonance imaging is a better exploration (4). Increased dialysis, especially daily dialysis, can reduce calcifications, but especially transplantation is the best treatment. Treatment of tumor calcinosis is sometimes surgical. While soft tissue calcifications in patients with ESRD is common, the prevalence of extensive periarticular cyst-like calcium salt containing space-consuming lesions in the form of tumoral calcinosis is very rare. We report a case of extensive TC occurring in an hemodialysis ESRD patient.

Case Report

A 61-year-old patient. He is followed, since 15 years, for high blood pressure complicated three years ago by hypertensive nephropathy for which he is under periodic hemodialysis, three times a week. He consults for mechanical pain in the left hip, the upper extremity of the right leg and both shoulders with swelling and limited range of motion in the right shoulder interfering with daily activity and evolving since three months. **The physical examination** showed a rounded mass at the anterior and supero-external right shoulder side, with regular contours, firm and mobile measuring 7 cm, along the deltoid muscle curve without local inflammatory skin signs (*Figure 1A*). In the right leg, the mass has the same characteristics, located on the supero-external side (*Figure 1B*). The mobility of knee was painful without limitation. The rest of the joint's exam, especially the two hips was normal. **Imaging examinations** revealed, at plain radiograph, severe manifestation of tumoral calcinosis: polycyclic, heterogeneous formations in the periarticular space of the 2 shoulders especially on the right one, voluminous and extending to the level of

the deltoid, the bursa under acromio-deltoid, supra-spinous and subscapular fossae, without bone erosion (*Figure 2A*). In the right knee, there is a multiple calcifications of the superior and postero-external soft tissue, at the proximal part of the internal collateral ligament, without focal bone lesion (*Figure 2B*). The MRI showed multiple macro-calcifications in the periarticular space of the 2 shoulders, especially on the right one. They occupy the synovium and peripheral muscles, in particular the deltoid, respecting the bone structures with fluid levels. These calcified formations are hypointense in T1, heterogeneous hypersignal with central areas of hyposignal in T2 (*Figures 2C and D*). **The blood test results** in admission found disturbance renal fonction : creatinemia at 94.30 mg / l, urea at 1.82 g / l. The calcium was normal at 103 mg / l with hyperphosphatemia at 71 mg / l, hyperparathyroidism at 128.50 pg / ml. Uric acid was at 69 mg / l. The aluminum level was normal. His calcitriol was at the lower reference range at 27 ng/ml. The final diagnosis of the patient was secondary TC due to ESRD with secondary hyperparathyroidism and hyperphosphatemia.

Due to the severity of the clinical and radiological findings, and because the patient refused at this stage any surgery, his hemodialysis schedule and number sessions have been increased, with low-calcium and low-phosphate diet advice. The follow-up radiography is expected in 3 months with control of laboratory examinations. The parathyroidectomy will be proposed if there will not be no response to this first treatment strategy.

Discussion

Tumoral calcinosis is a rare benign disease characterized by the deposition of calcium material in extra-articular soft tissues taking a tumoral form. It can be primary or secondary. Chronic renal failure in ESRD is one of the causes of metastatic calcifications (1,5). The prevalence of TC in hemodialysis patients has been reported to range from 0.5% to 1.9% (6).

The pathophysiology underlying the development of this disease entity remains unclear. It is mostly associated with manifestation of tertiary hyperparathyroidism, hypercalcemia and hyperphosphatemia. The other factors predisposing the patient to calcium salt deposits in outside of hyperparathyroidism and elevation of phosphocalcic product are: excess intake of calcium or vitamin D, poor compliance of the patient with regular intake of chelators, dialysis insufficient or a dialysate rich in aluminum (7).

Otherwise, the soft tissue calcification is considered secondary TC in patients with also pseudoxanthoma elasticum, malignancy, sarcoidosis, primary hyperparathyroidism, scleroderma, hypervitaminosis D, milk-alkali syndrome, or massive osteolysis (6). The periarticular calcifications may be accompanied by an aseptic inflammatory tissue reaction with encapsulation of the crystals. They are common on the hips, shoulders, knees, elbows, wrists and fingers. An homogeneous calcified pre-articular mass of soft tissues, with respect to the joint space without bone damage is the typical radiographic appearance (4).

Diagnosis of TC is mainly based on imaging modalities. Plain radiographs show the typical appearance of amorphous, multilobulated and cystic calcifications in a peri-articular location. The CT-Scan is useful

to appreciate the relationship of the tumor with the other organs just like the MRI which visualizes in addition the multiple cavities and the fluid levels. Single-photon emission tomography / CT hybrid imaging (PET-scan) mainly allows the identification of other subclinical joint or visceral locations in a single examination (6,8,9,10). Histology, when performed, find multiple lobules of calcified masses separated by septa of fibrous tissue surrounded by lymphocytes and giant cells (5).

The clinical context and the radiological data made the diagnosis of TC in our patient due to ESRD under dialysis. The rarity of this entity, the severity and the early onset of TC in secondary hyperparathyroidism state are the originality of this clinical case.

The global management is based on the correction of the calcium and phosphate product. The reduction or even the disappearance of calcifications go through the correction of hyperparathyroidism or the increase of dialysis sessions with low-Ca dialysate, while checking the aluminum level in the dialysate. Parathyroidectomy is one of the therapeutic options (11,12). Kidney transplantation can promote rapid decrease in calcifications and is indicate in cases of severe calcinosis. Surgery ,with complete resection, is recommended when there is a risk of vascular damage or on joint mobility (1,3,4).

Conclusion

We report a patient with ESRD under hemodialysis and severe tumoral calcinosis. It is important for rheumatologist and renal health care providers to recognize that ESRD patients may develop this rare disease entity to differentiate them to other causes of TC and in patient with ESRD to TC mimicking tophi. TC patients, including this case, should be checked regularly to keep their serum phosphate levels low, and radiologically assessed. The improvement of the treatment strategy and reinforcement of patient compliance are important for the complete remission.

Declarations

Disclosure of interests: None

Patient Consent: the patient has given his oral and written consent for publication of his clinical case.

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