

Images

Uremic tumoral calcinosis with atypical manifestation. Case report[☆]

Calcinosis tumoral urémica con manifestación atípica. Reporte de caso

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Tumoral calcinosis (TC) is a disease characterized by deposits of calcium phosphate crystals in soft tissues. The TC observed in patients with chronic kidney disease (CKD) is known as uremic tumoral calcinosis (UTC).¹ Below we present the case of a patient with terminal CKD, who presented calcification in the periarticular soft tissues of the fingers, which is an unusual manifestation of UTC.

This is a 48-year-old woman with terminal CKD due to polycystic kidney disease, on peritoneal dialysis for 3 years. The patient consulted for a 6-month history of symptoms which consisted of an increase in the size of the distal phalanx of the second and third fingers of the right hand, distal phalanx of the second finger of the left hand (Fig. 1) and a palpable mass on the right shoulder. The patient reported pain and itching at the site of the lesions, plus a non-fetid white discharge. The X-rays showed thick, amorphous calcifications of the periarticular soft tissues, compatible with TC (Fig. 2). The laboratory tests revealed tertiary hyperparathyroidism documented with parathyroid hormone values of 2362 pg/ml, albumin of 3.2 g/dl, phosphorus of 9.7 mg/dl and corrected calcium of 10.2 mg



Figure 1 – Photo of the patient's hand, with swelling in the distal phalanges of the fingers.

/dl. Parathyroid scintigraphy revealed hyperplasia of the 4 parathyroid glands. The patient underwent a parathyroidectomy of 3 glands, with a favorable evolution and a significant decrease in the lesions.

DOI of original article:

<https://doi.org/10.1016/j.nefro.2020.07.010>.

[☆] Please cite this article as: Aristizabal-Alzate A, Nieto-Ríos JF, Trujillo-Agudelo D, Cadavid-Aljure D, Serna-Higueta LM, Zuluaga-Valencia G. Calcinosis tumoral urémica con manifestación atípica. Reporte de caso. Nefrología. 2022;42:221–222.

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Figure 2 – X-ray of the patient's hands, with calcifications in the distal phalanges that had the hump.

TC is a rare disease, characterized by periarticular lesions of variable size, which involves small and large joints, leading to serious deformities, joint stiffness and functional impotence. They can also induce ulcerative lesions, allowing the transdermal elimination of calcific or milky material, as in the case described.²

TC is secondary to the presence of minimal repetitive trauma and a reparative inflammatory process. Apparently, trauma favors the calcinosis process by activating mechanisms that lead to hemorrhage, fat necrosis, fibrosis, collagenization, collagenolysis and, finally, calcification.³ Movement and friction are believed to be important in the formation of the adventitial bursa of periarticular location⁴; the bursa subsequently fills with calcified material, predominantly hydroxyapatite.

In the case described, calcinosis occurred in a patient with terminal CKD on peritoneal dialysis, who presented hyperparathyroidism with a marked alteration in phosphorus-calcium metabolism, and therefore we are dealing with a case of UTC. The prevalence of UTC in hemodialysis patients has been reported to be between 0.5-1.9%.⁵ CKD patients have several factors that can predispose to soft tissue calcification, the most important is an increase in the calcium-phosphorus product. The incidence of TC is high when the calcium-phosphorus product is greater than 55.⁶

Radiological evaluation has a key role in diagnosis. On plain radiographs, TC has an opaque and nodular appearance.⁷ Computed tomography and magnetic resonance imaging help to determine the extent of the lesions. Scintigraphy (methylene diphosphonate-technetium 99) is of great value in detecting multiple lesions, new lesions in formation, bone marrow involvement and to monitor therapy, since it reflects the activity of the lesions.

Most of the literature on TC describes that the location of lesions is around large joints.⁸ The specific manifestation presented in the case here described, with involvement of the distal phalanx of the fingers, generating a striking bulge, is not frequently described in the literature. Regarding the treatment of CTU, several strategies have been proposed, with preference

for medical management over surgical management. Since the most important pathogenic factor is the increase in the calcium-phosphorus product, the reduction of serum phosphorus represents a non-invasive and fundamental approach to therapy. Patients with hyperphosphatemia should receive phosphate binders. It has been suggested that daily dialysis with low calcium in the dialysate or daily dialysis at night could reduce TC.⁹ If hyperparathyroidism is present, parathyroidectomy has led to rapid resolution of lesions in some patients.¹⁰

Regarding surgical treatment, this is only used when the response to other treatments is insufficient. In the case described, tertiary hyperparathyroidism with hyperphosphatemia was demonstrated and it was decided to perform a parathyroidectomy. Surgical resection of the lesions was not considered in accordance with the recommendations in the literature. Post-surgical evolution was adequate, with improvement in calcinosis lesions in the patient's last follow-up.

In all uremic patients with periarticular calcifications, the diagnosis of TC should be considered; therefore, in these patients we recommend evaluating phosphorus-calcium metabolism, ruling out the presence of hyperphosphatemia and hyperparathyroidism, and treating these underlying conditions as the treatment of choice.

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