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Case Report

Pseudocalcinosis Tumorale (Teutschlander Disease) in Chronic Hemodialysis Patients

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Abstract

Pseudotumoral calcinosis (CPT) is a condition characterized by the deposition of calcium phosphate crystals in the periarticular tissues, forming large calcified masses. Although the pathophysiology of CPT is not fully understood, an increase in the calcium-phosphate product beyond the precipitation threshold and severe hyperparathyroidism appear to play a significant role. Treatment remains controversial, with surgical excision often recommended.

We report a case of CPT in a 74-year-old diabetic patient undergoing chronic hemodialysis who experienced progressively worsening pain in the left hip for six months, along with mobility difficulties. A CT scan revealed a calcified mass on the posterior thigh, likely explaining the electric shock-like pain, as well as compression of the superficial femoral artery causing decubitus pain resembling critical limb ischemia.

The biological assessment showed elevated calcium-phosphate levels and hyperparathyroidism. Surgical excision is not indicated due to the mass's proximity to vascular and nerve structures. This case highlights the diagnostic and therapeutic challenges of CPT, emphasizing the need for iterative angioplasties, considering that this condition is rare in chronic hemodialysis patients with calcified periarticular masses.

More Information

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Introduction

Calcified masses around the joints were initially identified and described by Duret in 1899 [1]. In 1935, Teutschländer [2] introduced the term lipocalcinogranulomatosis progressive, while Alberto, in 1943 [3], proposed the current name of tumoral calcinosis. These masses are often discovered with the appearance of a periarticular tumor affecting both small and large joints. They vary in size and can sometimes cause notable deformities, leading to joint stiffness and loss of function.

There are two clinical forms of this condition: the sporadic form, associated with chronic disease, and the familial form, likely genetic [4]. The sporadic form is common in patients with chronic renal failure, where calcium deposition is linked to disrupted phosphocalcic metabolism [5]. The familial form is thought to be inherited in an autosomal recessive manner, causing familial hyperphosphatemia due to enzymatic issues, without renal failure, often seen in African individuals with large periarticular masses in early childhood [6].

Our patient had a sporadic form due to chronic kidney failure. The exact pathophysiology of pseudotumoral calcinosis remains unclear, but some authors suggest that elevated phosphocalcium levels and severe hyperparathyroidism may be key factors [7]. Repeated joint trauma might also contribute. In dialysis patients, the prevalence of this condition ranges from 0.5% to 7%, likely due to hyperparathyroidism, a common complication of chronic kidney disease [8]. Excess parathyroid hormone promotes bone resorption, raising serum calcium and phosphorus levels, which may lead to soft tissue calcification [9].

The periarticular mass is usually painless, but there can be significant limitations in joint mobility depending on its location [10]. In our patient, the mass was located in the left thigh and indeed caused pain due to compression of the nearby sciatic nerve, leading to compromised mobility. While it may become symptomatic in some locations, causing pain through compression of surrounding structures.

The preoperative diagnosis is often not straightforward, presenting a challenge for surgeons to rule out a malignant tumor. The biological assessment is frequently normal [9]; however, disturbances in the phospho-calcium balance have been observed in this case.

Case report

A 74-year-old female patient, who is a chronic hemodialysis patient with type 2 diabetes, presents with critical limb ischemia. A mass is located on the left hip (Figure 1b), and another on the right leg (Figure 1a). The patient presents with decubitus pain, pulses are present and no skin lesions showed on clinical examination.

Elevated phosphate and calcium levels were noted, alongside impaired renal function with a creatinine level of 90.

The level of parathyroid hormone was recorded at 30 times the normal level.

Radiography showed juxtaposed articular calcifications that appear clustered due to small, dense, rounded images separated by radio-transparent septa (Figure 2).

Angioscanner shows high-density masses with pseudo-cysts and fibrous septa, often displaying the ''sedimentation sign'' (Figure 3a-c).

Discussion

In cases of chronic renal insufficiency, increased renal resorption of phosphate due to an abnormality of the vitamin D/PTH system or a primary anomaly of the proximal convoluted tubule of the kidney induces secondary hyperparathyroidism [11]. Over time, hyperphosphatemia and hyperparathyroidism lead to the formation of extracellular vesicles and secondary mineralization in the form of hydroxyapatite crystals. This is followed by the development of calcified peri-lesional granulomas in the form of tumoral calcinosis [12]. Other theories support a traumatic origin: McKee and Liomba favor dystrophic calcifications secondary to trauma that first leads to ischemia and then collagen damage, predisposing to calcifications [13].

The relationship between hyperparathyroidism and CPT is significant, as elevated parathyroid hormone (PTH) levels promote the mobilization of calcium from bone and increase renal tubular reabsorption of calcium. However, in

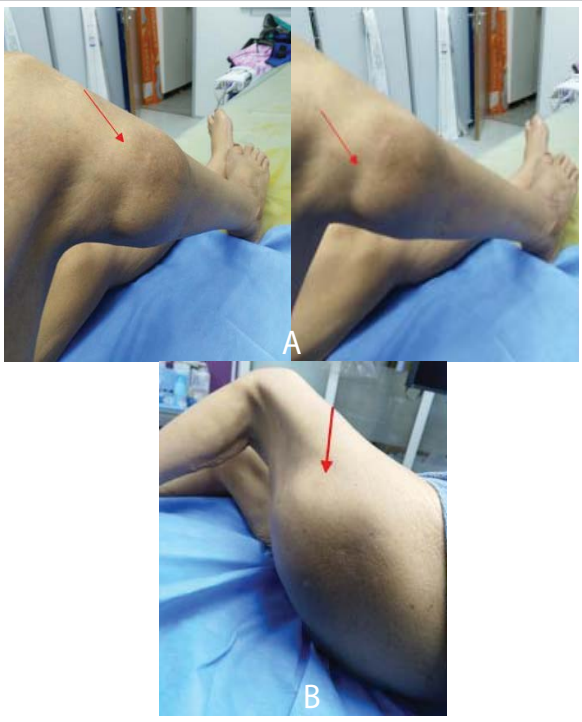

Figure 1: a: Mass on the lateral side of the right leg. b: Mass on the lateral side of the left thigh.

Figure 2: Calcified mass surrounding the left hip joint and upper thigh area.

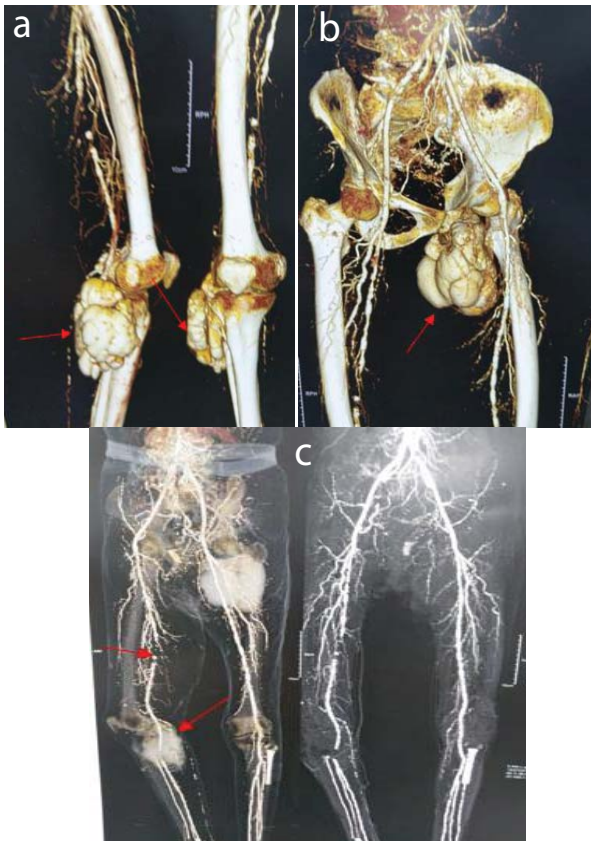

Figure 3: a: Masses in the left and right lower extremities. b: Mass in the left hip. c: Angioscan revealing occlusions of the superficial femoral artery and the stenosis of the popliteal artery above the joint.

chronic renal insufficiency, the kidneys are often unable to maintain normal serum calcium levels, leading to persistent hyperphosphatemia. This dual imbalance contributes to the ectopic calcifications characteristic of CPT, where hydroxyapatite crystals deposit in soft tissues, forming distinctive firm swellings [14].

Patients undergoing dialysis often experience dysregulation of calcium-phosphate metabolism due to their underlying renal dysfunction. Dialysis can manage fluid overload and waste products but may not adequately correct mineral imbalances. Although some phosphate is removed during dialysis sessions, dietary phosphate intake can lead to a net positive phosphate balance, exacerbating hyperphosphatemia and secondary hyperparathyroidism. Consequently, the interplay of elevated PTH and phosphate levels drives the pathogenesis of CPT, resulting in soft tissue calcifications even in patients compliant with phosphate binders [15].

The clinical presentation of CPT typically manifests as a firm, sometimes painful swelling that begins gradually, either singular or multiple. CPT can be complicated by skin fistulas and superinfections [16-18]. In the long term, there can be total functional impairment of the limb or even a picture of vascular and nerve compression in advanced forms [19]. The most common locations, in descending order, are the hip, elbow, shoulder, foot, leg, knee, and hand [20].

The standard first-line examination for diagnosing peri-articular masses is an X-ray, which can reveal juxtaposed articular calcifications appearing in clusters due to small dense rounded images separated by radiolucent septa [21,22]. These calcifications typically form a heterogeneous mass resembling a honeycomb structure [23].

Calculation of Ankle-Brachial Index (ABI) remains a diagnostic tool for detecting arteriopathy of the lower limbs. In this patient, symptomatology was often latent, despite the pseudocalcifying masses and repetitive compression of the arteries in the joint areas. This latency can be explained by the incompressibility of the arteries.

It is important to differentiate calcinosis from calciphylaxis, or "calcifying uremic arteriolopathy,". Calciphylaxis primarily affects patients with advanced renal disease undergoing dialysis and typically occurs about 30 months after the initiation of dialysis or even after kidney transplantation. It is characterized by calcifications in the media of small and medium-caliber vessels, as well as in the deep dermis and hypodermis, unlike calcinosis, which involves calcifications of soft tissues. Calciphylaxis leads to ulceronecrotic lesions, purpuric, livedoid, and hyperalgesic symptoms within the context of mineral and bone metabolism disorders, mainly hyperparathyroidism. The mortality rate is high: 60% to 80% in proximal locations and 20 to 30% in distal locations [24,25].

The clinical presentation of calciphylaxis is particularly concerning due to its association with significant morbidity and mortality. Patients often present with painful, necrotic skin lesions and systemic symptoms. The diagnosis of calciphylaxis requires a high index of suspicion, especially in patients with end-stage renal disease, and differentiating it from other causes of skin lesions is crucial for appropriate management [26].

The therapeutic modalities for uremic tumoral calcinosis are varied, though the results are sometimes insufficient. Stagnation of calcifications can be achieved in a few months by treating an established contributing cause: phosphorus restriction or the prescription of phosphate binders, along with stopping calcium and vitamin D intake, have shown some effectiveness and are considered more of an adjunctive treatment [27-29]. The efficacy of these measures is inconsistent, confirming the complexity of the pathophysiology. These therapeutic difficulties have led to the proposal of using a calcium-depleted dialysis bath "1.37 mmol/L" and especially to intensify the dialysis dose by increasing the weekly dialysis duration to more than 20 hours. Subtotal parathyroidectomy has yielded spectacular results in some patients, especially in cases of severe hyperparathyroidism. Indeed, Benkert et al., as well as Cohen and Parikh, observed complete regression with dissolution of tumoral calcinosis after subtotal parathyroidectomy [30,31]. For our patient, parathyroidectomy was not performed.

Endovascular treatment of critical ischemia, particularly when caused by this type of mass, is often palliative, especially in cases of functional impotence accompanied by persistent pain. In this patient, double dilatation with stenting of the superficial femoral artery, along with balloon angioplasty of the supra-articular popliteal artery, was performed to achieve revascularization of the lower limb. While this endovascular approach can provide symptom relief, it should not be considered curative and does carry a risk of rethrombosis [32].

Conclusion

Pseudotumoral calcinosis is a rare entity, which may be primary or secondary, and has a characteristic radiographic appearance on standard radiography, followed by CT. This condition can lead to critical ischemia as a result of mechanical compression by the calcifying mass, whose symptomatology may be latent due to the calcification of the vessels. The prognosis is generally favorable, but recurrence remains a real problem. Repetitive angioplasty (endovascular treatment) remains the most appropriate method, but its efficacy is limited in time due to recurrence.

Ethical considerations: Informed consent was obtained from the patient to publish the data.

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