Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Pseudo Tumoral Calcinosis and Renal Ostedystrophy in Pathien under Hemodialysis: About a Case

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DOI: 10.36347/sjmcr.2023.v11i10.035 | **Received:** 11.09.2023 | **Accepted:** 18.10.2023 | **Published:** 25.10.2023

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Abstract **Case Report**

Pseudo tumoral calcinosis is a rare affection, characterized by the deposition of calcium phosphate crystals in peri-articular soft tissues, forming giant mass, calcified and appearing as a compressive tumor. This affection can be hereditary of secondary to chronic renal failure undergoing hemodialysis. In this case, this affection is included in renal osteodystrophy. Radiology is the key for the diagnosis. We herein report the case of 68 years old, with chronic hemodialysis history, presenting with a latero-cervical mass, evolving since one year. CT showed diverse locations of pseudo tumoral calcinosis, associated to renal osteodystrophy and surgical resection was indicated for symptomatic locations.

Keywords: Hyperparathyroidism, chronic hemodialysis, pseudo-tumoral calcinosis, renal osteodystrophy, computed tomography.

INTRODUCTION

Pseudo tumoral calcinosis is secondary to the deposition of calcium phosphate crystals in extra-articular soft tissues, sometimes forming large calcified masses and compressing adjacent structures, particularly vascular and neural ones, depending on their location [1, 2]. Although the pathophysiology of pseudotumor calcinosis has not been fully elucidated, an increase in phosphocalcium above the precipitation threshold and severe hyperparathyroidism appear to play a major role [3]. In renal failure patients undergoing hemodialysis, the frequency of pseudotumoral calcinosis is estimated at between 0.5% and 7%, depending on the series [4].

OBSERVATION

We herein, report the case of a 58-year-old patient with a history of treated pulmonary tuberculosis, chronic hemodialysis for 10 years with three sessions per week, admitted to the university hospital center Mohammed VI of Oujda for management of a painless left laterocervical mass evolving for one year.

Clinical examination revealed a 5 cm long, firm, slightly mobile, painless, roughly oval, left latero-cervical formation with no local inflammatory signs. General examination also revealed the presence of other formations with the same characteristics as the one described above on both shoulders and in the posterior gluteal region opposite the left hip joint. Biological findings included a hyperphosphoremia with hypocalcaemia and hypercalciuria, at and elevated parathyroid hormone.

A cervico-thoraco-abdomino-pelvic CT scan without and with injection of contrast agent was performed, revealing the presence of:

- Multiple calcified and multiloculate masses and nodules in the periarticular soft tissues. In case of voluminous mass, a liquid-calcification level can be seen. These masse affected the soft tissues of the left latero-cervical region, the shoulders and the left hip (Figures 1, 2 and 3).
- Staged spinal osteocondensations appearing as "Rugby shirt" appearance, with multiple microlytic lesions of the skull bones giving a salt-and-pepper sigh appearance in connection with renal ostodystrophy (figure 4).

Surgical removal of symptomatic calcinosis of the left cervical region and left shoulder, together with parathyroidectomy and medical treatment are indicated.

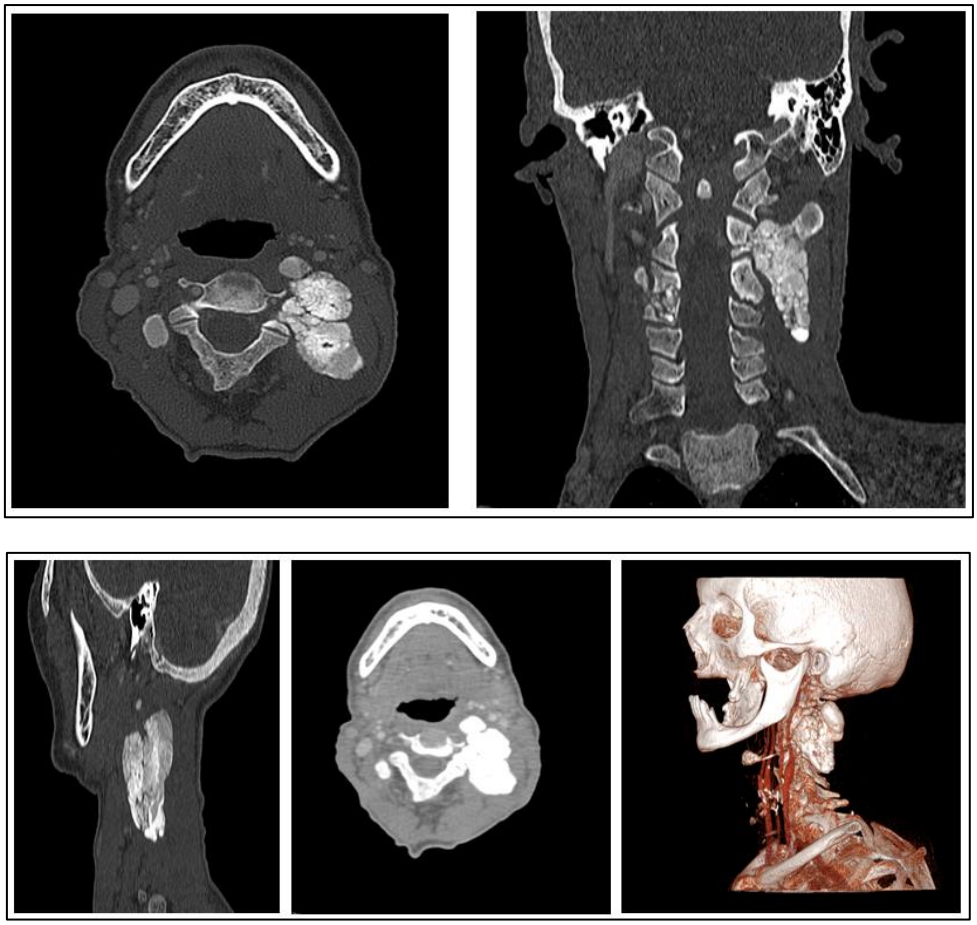

Figure 1: cervical CT scan in axial, coronal and sagittal planes, and in 3D reconstruction, with and without enhacenemnt showing a left latero-cervical voluminous mass, which extends from C1 to C4, over a length of 5 cm, with an endoforaminal extension C2-C3, and compresses the vertebral artery at this level which remains permeable.

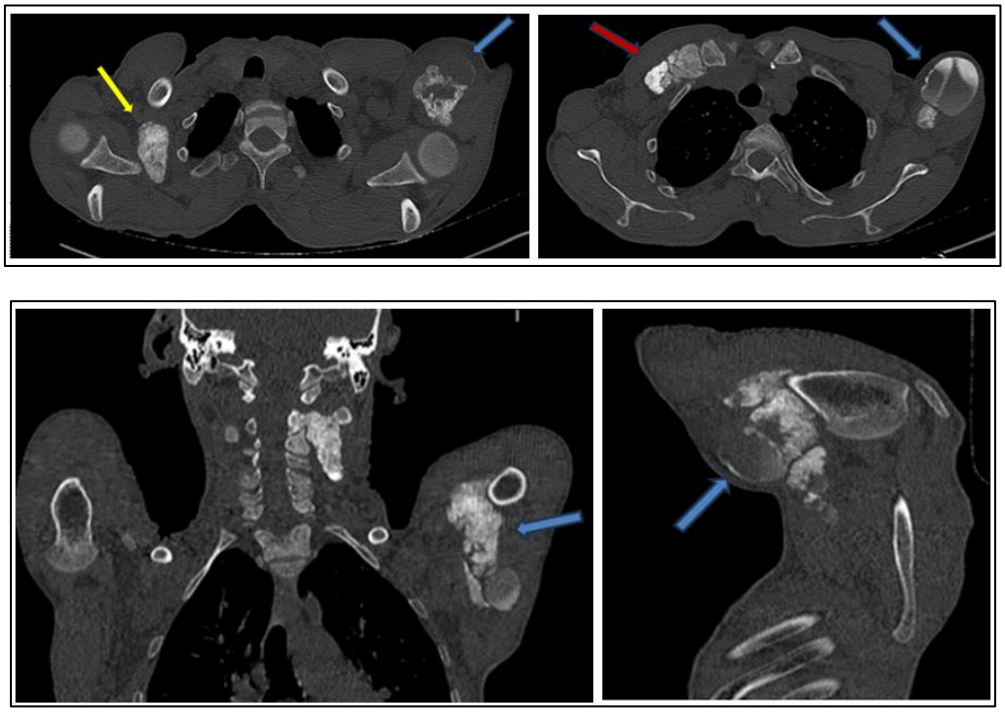

Figure 2: Cervicothoracic CT scan in bone tissues window, in 3 different planes showing 3 formations in the left shoulder (blue arrow), the right shoulder(yellow arrow) and near the right sterno-chndro-clavicular joint(red arrow), totally calcified on the right and partially calcified on the left, containing a liquid-liquid level associated with sedimentation deposits.

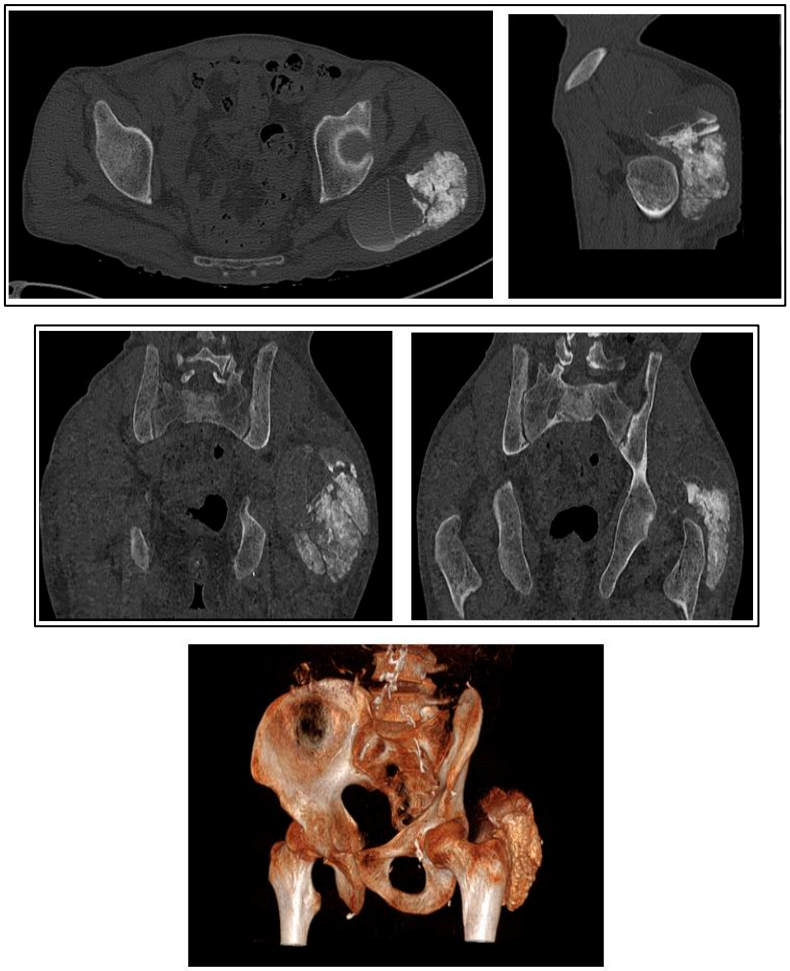

Figure 3: CT scan of the left hip in axial, sagittal and coronal planes, and in 3D reconstruction showing a 125 mm long pseudotumoral calcinosis mass, partially calcified.

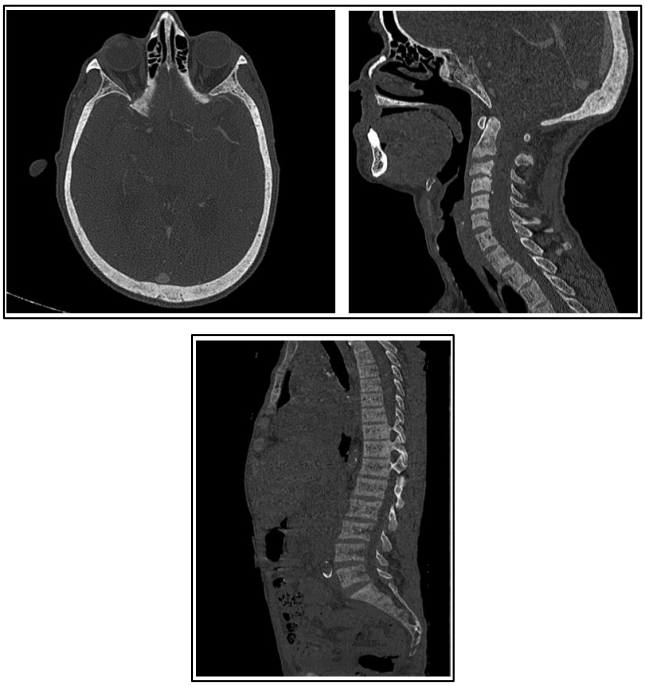

Figure 4: CT scan of the brain and the entire spine in bone window showing osteocondensation of the plateaus of the vertebral bodies of the entire spine, giving a "rugby shirt spine" appearance, with multiple microlytic lesions of the skull bones, giving a salt-and-pepper appearance related to renal osteodystrophy.

DISCUSSION

General

Pseudotumoral calcinosis is a rare affection characterized by calcific deposit in periarticular soft tissues [1]. It has been described under various names since the early 1900s, but the current name was coined by Inclan *et al*., in 1943 [5]. Just over 200 cases have been reported in the literature [6].

Etiopathogenesis

There are two clinical forms:

- The familial form, called primitive, probably of genetic origin due to autosomal recessive inheritance, and is responsible for familial hyperphosphatemia by enzyme dysregulation without necessary renal failure associated. According to some authors, it is the prerogative of the African subject with voluminous periarticular masses during the first decade of life [7].
- The sporadic form, secondary to chronic disease [5, 6], is most often found in people with chronic renal deficiency, and in these cases, calcium deposition is linked to an abnormality in phosphocalcic metabolism [3].

Repeated joint trauma may also play a role. In dialysis patients, the frequency of pseudotumoral calcinosis is estimated at between 0.5% and 7%, depending on the series. This could be explained by the effects of hyperparathyroidism, an unavoidable complication of chronic renal failure. Excessive parathyroid hormone stimulates bone resorption, thus increasing serum calcium and phosphorus levels, which in turn promote the deposition of crystals in soft tissues, which may explain the long-term manifestations of pseudotumoral calcinosis [8].

In our case, it was a secondary pseudotumoral calcinosis in a patient with chronic renal failure under haemodialysis since 10 years complicated by hyperparathyroidism.

Clinical Features

In the majority of cases, calcinosis pseudotumoris presents as a firm, painless swelling, mobile, with variable size but often voluminous and responsible for compression of adjacent structures, particularly nerve and vascular structures. The most common sites are the hip, shoulder, elbow, sacrum and ankle [9].

Biology

In the majority of cases, serum calcium, calciuria, phosphaturia and alkaline phosphatase are normal; serum phosphatase is usually elevated.

Imaging: [10- 12].

- Standard radiography: shows the presence of plurilocular para-articular lesions with juxtaposition of "clusters" of dense, amorphous, well-limited calcifications.
- Ultrasound: the sonographic appearance varies according to calcific density: attenuating hyperechoic mass or presence of cavities with a liquid-liquid level.

- **Computed Tomography (CT):**

It shows multilocular periarticular masses with juxtaposition of dense, amorphous, well-limited calcifications showing as “clusters”, sometimes containing sedimentation as a liquid-calcification level, reflecting the sedimentation of calcium crystals in a serous supernatant. Granulomatous reaction may cause erosion of the cortical bone on contact.

It determines the local extension and relationship with adjacent structures, and is used to assess distant extension.

It can also identify other osteoarticular lesions associated with renal osteodystrophy.

- **Magnetic Resonance Imaging (MRI):**

Calcified masses are well limited, appearing hyposignal inT1 weighted image, variable signal in T2 weighted image depending on the amount of calcium and serous supernatant (liquid-liquid level)

- **Nuclear Medicine:**

Allows early detection of periarticular calcifications due to biphosphonate binding, and to demonstrate hypermetabolism if local inflammation by 18FDG PET.

Treatment

Complete surgical excision is the rule for any symptomatic pseudotumoral calcinosis due to its volume and/or location [3]. Surgical treatment consists of complete removal of the lesion to ensure that the tumor does not recur. Incomplete removal of the lesion will lead to tumor recurrence.

Post-operative evolution is generally straightforward, and the prognosis remains good. [3]. Nevertheless, the recurrence rate is high, especially after incomplete resection and in patients with secondary hyperparathyroidism [13]. Medical treatment phosphorus binders can be used, and treatment of hyperparathyroidism and renal failure are necessary.

CONCLUSION

Pseudotumoral calcinosis is a clinical and histological entity involving the deposition of calcium phosphate crystals in periarticular soft tissues, which may be primary or secondary, particularly in cases of

chronic renal failure. Imaging plays an important role for the diagnosis, the extension assessment and the follow-up of tumoral calcinosis after appropriate treatment.

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