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Case Report

Tumoral Calcinosis in a Patient on Hemodialysis Involving Cervical Spine and Hips

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Abstract

A 58-year-old white man with chronic renal failure, presented with tumoral calcinosis involving cervical spine, and both hips, manifesting as severe upper cervical pain, and hip movement limitation. CT scan revealed a periarticular calcified mass with no bone involvement at the level of the vertebrae cervical, and two calcified masses in both hips. The diagnosis was made based on the radiological appearance and subsequently confirmed by anatomopathological examination. Tumoral calcinosis is a rare benign disease, characterized by calcified deposits in periarticular tissues. Only five cases of tumoral calcinosis of the upper cervical spine have been reported in the literature, our case is the sixth. The pathogenesis is yet not well understood. Surgery remains the treatment of choice, and the prognosis is generally favorable. We aim to underline the role of radiology in diagnosing this entity with an accompanied review of the literature.

Keywords: *Tumoral Calcinosis; Cervical Spine; Hips; CT Scan; Hemodialysis*

Introduction

Tumoral calcinosis is a rare hereditary phosphate regulation dysfunction associated with the formation of massive periarticular calcinosis in extracapsular soft tissues. Usually discovered around large joints, such as the hip, elbow, and shoulder, and can be accompanied by palpable mass near the affected joint [1]. Tumoral calcinosis of the vertebrae cervical is extremely rare. Only five cases of tumoral calcinosis of the upper cervical spine have been reported in the literature, our case is the sixth. We report the case of a 58-year-old white man with chronic renal failure, presented with tumoral calcinosis involving the vertebrae cervical, and both hips, manifesting as severe upper cervical pain, and hip movement limitation. CT scan revealed a periarticular calcified mass with no bone involvement at the level of the vertebrae cervical, and two calcified masses in both hips. The diagnosis was made based on the radiological appearance and subsequently confirmed by anatomopathological examination. We aim to underline the role of radiology in diagnosing this entity with an accompanied review of the literature.

Case Report

A 58-year-old white man with a 26-year history of chronic renal failure and receiving hemodialysis three times a week was admitted with a main complaint of hip movement limitation, with a palpable mass at the level of the right and left hips of progressive evolution over

5 years, the evolution was characterized by the installation of neck rigidity with severe upper cervical pain, spastic gait, and weakness of the left upper extremity since one year. Physical examination revealed a firm, hard mass in both hips and the upper left cervical region, with limitation of hip movement more marked on the right, with atrophy and weakness of the musculature of the lower limbs, and the left upper limb, the rest of the physical examination being within normal limits. Initial laboratory tests of serum calcium, alkaline phosphatase, and uric acid levels found values within the normal limits and the C-reactive protein test was negative. Hemodialysis controlled the blood urea nitrogen and serum creatinine levels to within 50 mg/dl (normal value: 15 - 40 mg/dl) and 5 mg/dl (normal value: 0.70 - 1.3 mg/dl), respectively. The patient is referred to our radiology department for a CT scan of the cervical spine and pelvis. At the level of the cervical spine (Figure 1), we note the presence of a periarticular calcified mass of the left cervical vertebrae, extending from C2 to C4, well-limited, with polylobed contours, made of amorphous calcifications, insinuating at the level of the transverse foramen without endocanal extension. At the level of the pelvis (Figure 2 and 3), we noted the presence of two periarticular calcified masses at the posterior area in both hips, with the right mass extending to the upper third of the thigh. The diagnosis of tumoral calcinosis was made based on the radiological appearance and subsequently confirmed by anatomopathological examination.

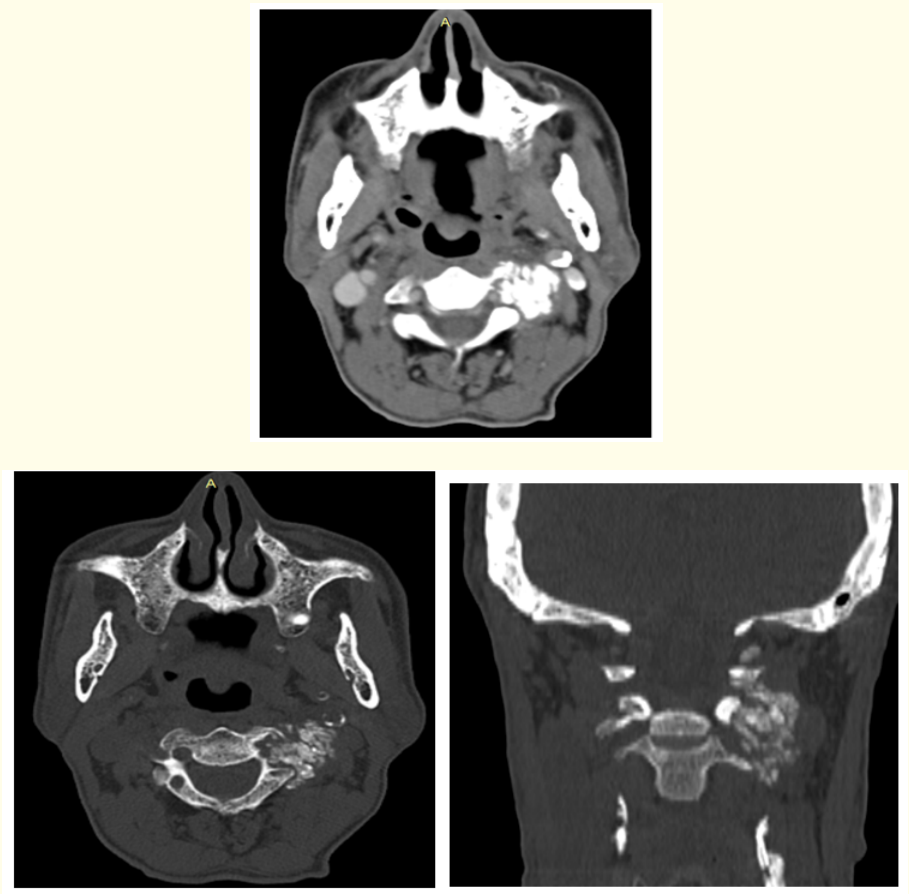

Figure 1: *CT scan of the cervical spine, showing a periarticular calcified mass of the left cervical vertebrae, extending from C2 to C4, well-limited, with polylobed contours, made of amorphous calcifications, insinuating at the level of the transverse foramens without endocanal extension.*

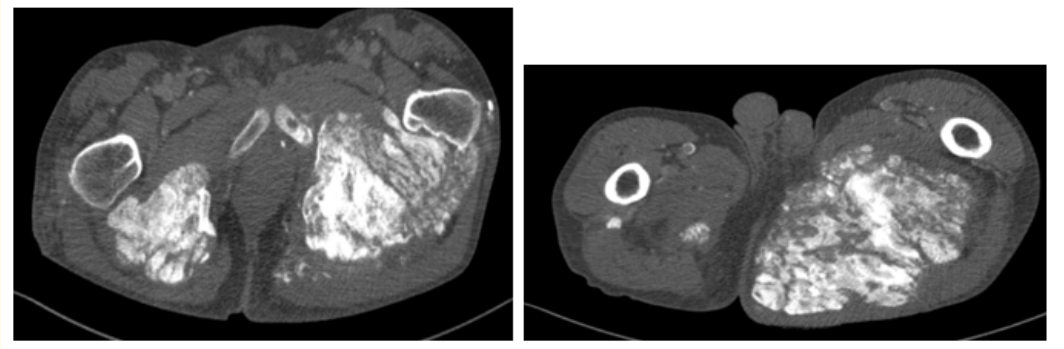

Figure 2: *CT scan axial images of the pelvis demonstrate two periarticular calcified masses at the posterior area in the both hips, with the right mass extending to the upper third of the thigh.*

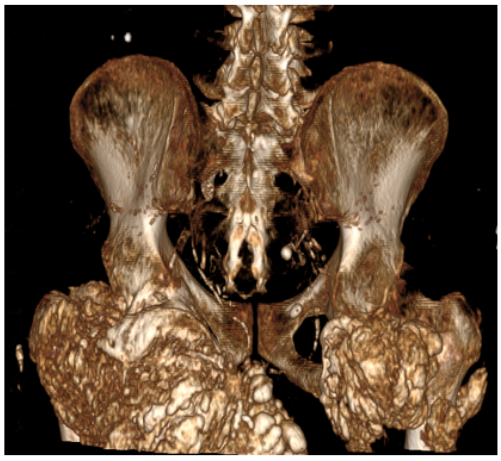

Figure 3: *Tridimensional CT scan demonstrates two calcified masses in both hip.*

Discussion

Tumoral calcinosis is a rare disease characterized by calcium deposits in the periarticular soft tissues [2]. This disease has had various names since the early 1900s, but Inclan., *et al*. in 1943 gave us the present name [3]. A little more than 200 cases have been documented in the literature [4]. Tumoral calcinosis is most common in teenagers and young adults of the black race [5,6].

There are three types of tumoral calcinosis: (a) an autosomal recessive mutation in one of three genes (fibroblast growth factor (FGF) 23, KL, and GALTN3) that causes phosphate metabolism errors, (b) a complication of renal failure and dialysis (as in our case), and (c) a sporadic incidence [7].

Clinically, it appears as a slow-growing soft tissue mass, non-erythematous, painless in most cases, arising near large joints, mobile regarding the skin but adherent to the deep plane without the involvement of the osseous elements themselves [1,7,8]. The size varies, and there are numerous localizations, sometimes bilaterally and symmetrically. The hip, shoulder, elbow, sacrum, and foot are the most commonly affected areas [7,9,10]. Only five cases of tumoral calcinosis of the upper cervical spine have been reported in the literature, two adults presented with severe neck pain, one with progressive radiculomyelopathy, and two children presented with torticollis [11-15]. our case is the sixth. The pathogenesis of these pseudotumor lesions is unclear [7,9].

Tumoral calcinosis shows on X-rays as a well-defined periarticular calcified mass of soft tissue, typically on the extensor side of the articulation. The multilobulated aspect is composed of numerous round or oval opacities of varying sizes and densities. The lobules are divided by radiolucent lines that correspond to the fibrous septum histologically [1,16]. On a CT scan, it may be composed primarily of large cystic components with low attenuation centers and thin layers of calcium outlining the walls. A more nodular calcified component is more frequently present, separated by low-attenuation septations. Some septa may enhance after contrast injection. A CT scan reveals no bone involvement and allows the detection of other bone anomalies [1,16,17]. On MRI, it appears as a well-circumscribed multicystic mass, with an intermediate inhomogeneous signal on T1-weighted images. heterogeneous hyper-signal on T2-weighted images, despite the large calcium component. A low signal of the entire lesion in all sequences has been described [1,5,16]. The septa separating the cysts are hypo-signal on T1- weighted images, a variable signal on T2-weighted images, and enhance after Gadolinium injection. The inner layers of the septa can hold calcified incrustations, which explain the low signal on both T1- and T2-weighted images. The outer layers are composed of connective tissue associated with a variable degree of vascularization and inflammatory reaction, accounting for the high intensity present on T2-weighted and post-contrast T1-weighted images [16]. On ultrasound, when the lesion is not highly calcified, it presents as a multiloculated mass with multiple cavities bounded by thin echoid septa. Some of these septa may be vascularized in Doppler color. The cavities are filled with anechogenic or echogenic fluid. When the lesions are entirely calcified, they appear as a hyperechoic mass with an acoustic shadow [5,16,17].

Hemangioma or calcified lymphangioma, teratoma, parosteal osteosarcoma, hypervitaminosis D, and other conditions are considered in the differential diagnosis of tumor calcinosis due to chronic renal failure. These diseases are excluded from the clinical context of chronic renal failure, and the laboratory results in the case of our patient, the pathological examination had the final say in confirming the diagnosis [5].

The majority of the time, treatment is symptomatic and palliative. Although surgical removal is a common choice, there have been few reports of spontaneous removal. In contrast, inadequate excision leads to a high rate of recurrence in patients with and without metabolic disturbances, and the development of recurrent masses is more frequently rapid than that of the initial lesions [18].

Conclusion

Tumoral calcinosis is an uncommon disease that can be secondary to chronic renal failure. It has a specific radiographic appearance on conventional radiography, CT scan, and MRI. Surgery remains the treatment of choice, and the prognosis is generally favorable.

Conflict of Interest

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Author's Contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

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