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## Case Report

# The idiopathic localized tumoral calcinosis: the “chicken wire” radiographic pattern

Marco Di Serafino MD<sup>a,\*</sup>, Matilde Gioioso MD<sup>a</sup>, Rosa Severino MD<sup>a</sup>, Francesco Lisanti MD<sup>b</sup>,  
Rosario Rocca MD<sup>a</sup>, Paolo Sorbo MD<sup>c</sup>, Domenico Maroscia MD<sup>a</sup>

<sup>a</sup> Emergency Radiology Department, San Carlo Hospital, Potito Petrone St, Potenza 85100, Italy

<sup>b</sup> Emergency Department, San Carlo Hospital, Potenza, Italy

<sup>c</sup> Surgery Department, Luccioni Clinic, Potenza, Italy

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## ABSTRACT

Tumoral calcinosis is a rare and benign hereditary tumor-like periarticular calcium deposit. It is painless and it is found commonly around large joints such as hip, shoulder and elbow. The condition predominately affects young black African patients with an equal gender ratio. In this report, a case of primary idiopathic localized tumoral calcinosis in a 22-year-old African man and its distinctive radiographic “chicken wire” pattern was described.

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## Case report

A 22-year-old African man was presented with an isolated left elbow, not datable, mass restricted joint movement (Fig. 1). The mass was painless, mobile beneath the skin and subcutaneous tissues, with hard consistency and lobulated. The clinical history was not helpful and no other significant symptoms were referred. A family-like condition was suspected by his medical history. Biochemical investigations were not relevant: serum calcium, parathyroid hormones, renal function, serum alkaline phosphatase and other biochemical tests were normal. X-rays (Fig. 2) revealed a calcified subcutaneous and periarticular left elbow mass with no bone reaction. The lobulated appearance and the

conglomeration of multiple small opacities separated from each other by thin linear radiolucent lines like a “chicken wire” pattern were suggestive of tumoral calcinosis. Computed tomography (CT) scan (Fig. 3) was performed for preoperative assessment of the large mass. At surgery, the whole tumor was removed (Fig. 4) with a less clear plane of separation from surrounded structures firmly attracted to the underlying fascia. The study of the gross specimen revealed a large subcutaneous, not vascularized, calcified mass, well-circumscribed, although not encapsulated, with the nodular surface (Fig. 5) measuring 10 cm of diameter. Histological study of the section confirmed a tumoral calcinosis in an inactive metabolic phase. The postoperative course was free from complications without local or other sites recurrences.

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\* Corresponding author.

E-mail address: [marcodiserafino@hotmail.it](mailto:marcodiserafino@hotmail.it) (M. Di Serafino).

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**Fig. 1 – Isolated left elbow subcutaneous mass (arrows).**

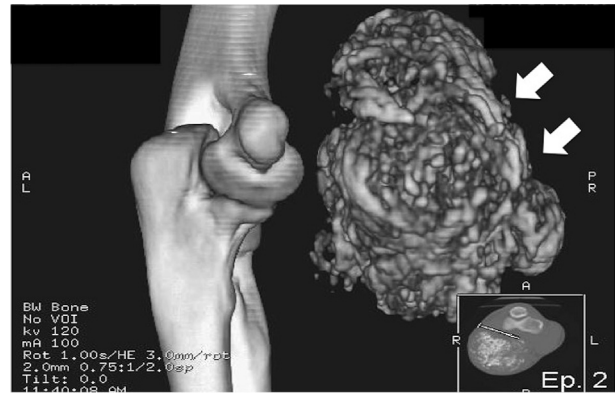
## Discussion

Idiopathic tumoral calcinosis is an unusual benign condition characterized by the presence of calcified soft tissue masses of varying size around the joints [1]. The term “tumoral calcinosis” was first stated by Inclan et al [2] in 1943 in the American literature for a disease characterized by large juxta-articular lobular calcified masses without visceral or skin calcifications in patients showing normal serum calcium and phosphorus levels [3]. The disease consists in a rare benign tumor-like, periarticular calcium deposit and is more often multiple and bilateral in the elbow, hip and shoulder of black race, healthy children and young adults of the same family [4]. The hereditary condition associated with massive periarticular calcification is the key point of this tumoral condition [5].

The tumoral calcinosis is a clinical-pathological entity that is rarely observed in the European population and North America, whereas it is more common in African populations [6,7].



**Fig. 2 – X-ray lateral projection of the left elbow shows periarticular well-circumscribed and multilobular area of multiple small opacities (arrows) separated from each other by thin linear radiolucent lines like a “chicken wire” pattern. The underlying bone is normal in appearance.**



**Fig. 3 – CT—volume rendering 3D sagittal view reconstruction shows a mass of increase density in the soft tissue anterior to the elbow joint (arrows).**

The tumoral calcinosis is often characterized by multiple rather than solitary periarticular pseudotumoral calcium deposits predominantly localized in the regions of the shoulder, elbow, hip, foot and wrist [8,9]. The disease is mostly found in children, adolescents and young adults [8]. From the viewpoint of the etiological factors, they are considered as genetic disorders, microtrauma, kidney failures, and calcium and phosphorus metabolism disorders. [9] In the past, they have also drawn the chromosome and the gene involved in the pathogenesis of familial form of tumoral calcinosis [10]. In fact, mutations of the gene coding for the protein SAMD9 [11] have been discovered, as well as mutations in the gene GALNT3 and the Klotho gene too [10–12]. There are two distinct phases of the disease: an active and an inactive phase, which can coexist in the same lesion. In the active phase or cellular phase, the calcified amorphous material, contained inside cystic spaces, appears surrounded by a proliferation of histiocytes giant cells and long-term inflammatory elements [8]. In the inactive phase, it shows only calcified material, surrounded by a dense ipo-cellular fibrous tissue that extends to adjacent structures [8]. Slaviv et al [13] have proposed a classification of tumoral calcinosis in 3 stages: the stage 1 with a proliferative lesion without calcifications; the stage 2 with a cystic lesion and calcifications; the stage 3 with an inactive calcified ipo-cellular lesion liken to this study's case. Some calcified periarticular lesions similar to those of tumoral calcinosis, can be observed in patients with long-term renal failure and secondary hyperparathyroidism, although these patients are older than those with tumoral calcinosis [14]. In addition, it can also be observed in patients with hypervitaminosis D, hyperparathyroidism, milk-alkali syndrome and in patients with massive osteolysis for inflammatory or neoplastic diseases. However, the latter conditions are accompanied by a condition of hypercalcemia [8–15]. Tumoral calcinosis may also be associated with scleroderma and polymyositis [16]. Consequently, the secondary calcified lesions to the above pathologies must be differentiated from calcinosis tumor. The soft-tissue lesions of tumoral calcinosis are typically lobulated, well-demarcated calcifications liken to this study's case. The diagnosis of tumoral calcinosis is mainly based on the typical characteristics of imaging (radiography and CT) and biochemical profile. Tumoral calcinosis has a

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