

Ayak bileğinde lokalize tümoral kalsinozis olgusu

A case of tumoral calcinosis localized on the ankle

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Abstract

Calcinosis cutis is a term used to describe a group of disorders in which insoluble compounds of calcium are deposited within the skin due to local and/or systemic factors. Cutaneous calcification may be divided into four major categories; dystrophic, metastatic, idiopathic and iatrogenic. Tumoral calcinosis is a rare disease that is a subgroup of idiopathic calcinosis cutis with an obscure pathology characterized by the deposition of calcium in periarticular areas mainly around the hip, shoulder and elbow. Here we present a 61-year-old woman presented with a 3- month history of an asymptomatic nodular lesion growing in time on the left ankle that was diagnosed as tumoral calcinosis with the clinical, histological and radiographic findings.

Key words: Calcinosis, calcium, cutaneous

Özet

Kalsinozis kutis; lokal veya sistemik faktörler nedeniyle, çözünmeyen kalsiyum bileşiklerinin depolandığı bir grup hastalığı ifade etmek için kullanılan bir terimdir. Kutanöz kalsifikasyon dört ana sınıfa ayrılabilir; distrofik, metastatik, idiyopatik ve iyatrojenik. Tümoral kalsinozis, esasen kalça, omuzlar ve dirsekte olmak üzere periartiküler alanlarda kalsiyum depolanması ile karakterize, idiyopatik kalsinozisin bir alt grubunu oluşturan nadir bir hastalıktır. Burada 61 yaşında, sol ayak bileğinde 3 aylık asemptomatik nodüler lezyonu olan, klinik, histolojik ve radyografik bulgularla tümoral kalsinozis tanısı almış bir kadın hasta sunmaktayız.

Anahtar kelimeler: Kalsinoz, kalsiyum, kutanöz

Introduction

Calcinosis cutis is an uncommon disorder and defined as deposition of insoluble calcium salts in cutaneous tissues (1). Cutaneous calcification may be divided into four major categories: dystrophic, metastatic, idiopathic and iatrogenic (1). There is no underlying disease in idiopathic calcinosis cutis (1-4). Tumoral calcinosis is a rare disease that is a subgroup of idiopathic calcinosis cutis and

characterized by the deposition calcific masses around major joints such as hips, shoulders, elbows and knees (1,2). We report a case of tumoral calcinosis in an adult localized on the ankle.

Case

A 61-year-old woman presented with a 3- month history of an asymptomatic nodular lesion growing in time on the left ankle. There was no history of trauma. Physical examination revealed a hard, skin-coloured,

sharply circumscribed lobulated nodular lesion, 4 cm in diameter on the left ankle lateral malleol (Fig. 1). All laboratory investigations, which include serum glucose, serum calcium (ionized and free) and phosphate levels, renal, liver function tests, hormone levels (thyroid hormone, parathyroid hormone, vitamin D) were within normal limits. Autoimmune markers (ANA, anti-dsDNA) were negative. Urinary calcium and phosphate levels were also in normal ranges. The direct radiographic examination of the left ankle showed soft tissue calcification in a 'wet cotton ball' pattern consisted with the calcium deposition (Figs. 2a and 2b). In histologic examination of the totally excised material with routine hematoxylin and eosin stain, epidermis was hyperkeratotic and dermis was widely infiltrated by deposits of calcific material (Fig. 3). There was no cartilage, bone formation or atypical cell. The case was diagnosed as tumoral calcinosis with the clinical, histological and radiographic findings.

Discussion:

Idiopathic calcinosis cutis occurs without identifiable underlying tissue abnormalities or abnormal calcium and/or phosphate metabolism (1,2,3,4). Idiopathic calcification includes four subgroups: idiopathic calcification of the scrotum, tumoral calcinosis, subepidermal calcified nodule and milia-like idiopathic calcinosis cutis in Down syndrome (5). In 1899 Duret observed a process of calcification in siblings and found this condition as an entity, in 1943 the term tumoral calcinosis was introduced by Inclan (6,7,8). The same condition has also been reported under the names of 'lipocalcinogranulomatosis' and 'calcifying collagenolysis' (7,8).

Tumoral calcinosis is a disorder with an obscure pathology characterized by the deposition of calcium in periarticular areas mainly around the hip, shoulder and elbow (7,9). The disease occurs

in three clinical settings (10). First, idiopathic or normophosphatemic form that is seen in young adults, primarily in African natives that is not familial and lesions are usually solitary and antecedent trauma is frequently present (10). These patients have no known calcium or phosphorus abnormality (10). Second, primary hyperphosphatemic tumoral calcinosis is a familial disease, primarily of black males in the first or second decades of life and manifested as hyperphosphatemia, elevated serum 1,25-dihydroxyvitamin D, normal serum calcium level (8,10). It is believed to be inherited according to a dominant autosomal pattern and has occasionally been found in siblings (8,10). Third, tumoral calcinosis is a potential complication of renal dialysis (10). In renal failure patients tumoral calcinosis is observed as a result of secondary hyperparathyroidism (10).

The lesions of tumoral calcinosis are characterized clinically by the presence of irregular, painless and periarticular soft tissue calcifying masses (9). In radiological findings striking signs are the cloud-like calcification structures which are polycyclically circumscribed (10). Histopathologic examination shows deposition of amorphous substances with evidence of calcification associated with a foreign body granulomatous reaction (10). The clinical presentation, radiological and histopathological features of our case was typical for tumoral calcinosis. The patient with solitary painless lesion on the left ankle was free of any renal, metabolic diseases. She had no familial history. On the basis of these findings the case is diagnosed as form of normophosphatemic tumoral calcinosis.

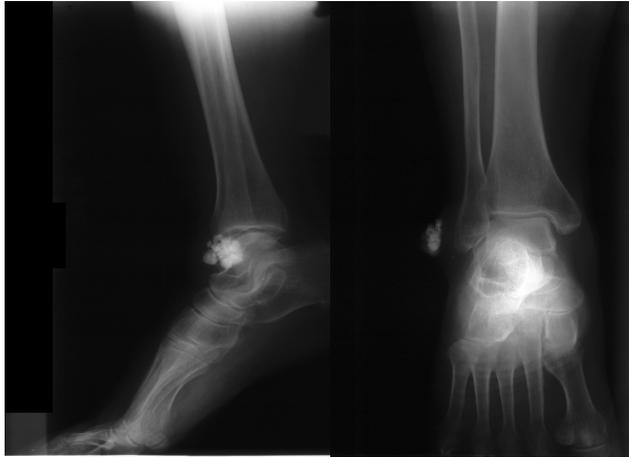
Differential diagnosis of the disease includes other forms of calcinosis cutis, cartilage lesions and osteoma cutis. As the patient's serum calcium and phosphate levels were normal, she had no functional parathyroid disorder and metabolic abnormalities and had no signs of Raynaud's phenomenon, skin

disease or telangiectasia. The plain radiographic features of a bone-forming lesion or cartilage calcification were not present. In histologic examination there was no cartilage or bone formation in any of the lesions.

Surgical excision has been the mainstay of therapy for tumoral calcinosis (8, 9). There is a high incidence of recurrence, especially after

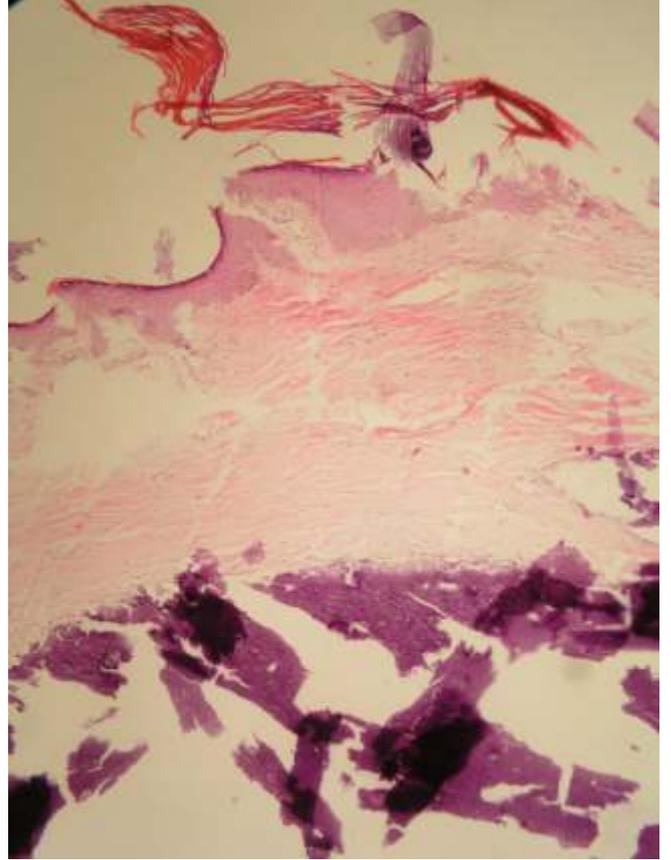


Resim 1: Clinical appearance showing hard, circumscribed lobulated nodular lesion on the ankle



Resim 2: Anteroposterior and lateral radiograph of the left ankle. Pronounced calcification structure on the left ankle, typically circumscribed.

incomplete excision and in patients with secondary hyperparathyroidism (6). Our patient was treated by totally excision and there has been no sign of recurrence one year later. Since tumoral calcinosis is diagnosed rarely, we find this case noteworthy to be presented with its typical clinical presentation.



Resim 3: Histology of excised nodule showing calcium deposits in dermis (H&E).

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