

Extensive Tumoral Calcinosis in a Patient With Systemic Sclerosis

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Abstract

Tumoral calcinosis, a rare disease manifesting with periarticular, calcified masses in soft tissues, can be either primary or secondary to a disease, such as systemic sclerosis. In the case reported here, a patient diagnosed with systemic sclerosis presented with hard calcified masses that involved the shoulders and hips.

In this article, we report the rare case of a woman who had scleroderma and presented with extensive calcified soft-tissue masses in both her hip and shoulder joints. The term *tumoral calcinosis* is used to describe tumor-like calcified masses occurring adjacent to large joints, including hip, shoulder, and elbow. It can be a primary, hereditary condition, or a condition secondary to various clinical entities, such as collagen diseases. In our patient, the extent of calcifications was remarkable; they involved numerous large joints. We could not find a report of a similar case in the literature. We discuss tumoral calcinosis related to scleroderma and briefly review the literature. We have obtained the written informed consent of the patient for print and electronic publication.

CASE REPORT

A 68-year-old white woman was referred to us with a 6-month history of pain in both her hips and shoulders. No history of trauma was recalled. She had been diagnosed with scleroderma 15 years earlier, when she presented with Raynaud phenomenon and symmetrical skin thickening involving the trunk and limbs. Clinical and laboratory findings at that time were consistent with the diagnosis of scleroderma.

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When the patient was referred to us, she was being treated with methotrexate, piroxicam, and omeprazole. Physical examination revealed subtle limitation of motion and mild tenderness over the affected joints. In both hip joints, there were firm, plaque-like, soft-tissue nodules. The patient had no family history of tumoral calcinosis. Neurologic examination findings were normal. At time of presentation, laboratory values were 9.5 mg/dL serum calcium (reference range, 8.0 to 10.5 mg/dL), 0.9 mg/dL serum creatinine (reference range, 0.6 to 1.3 mg/dL), 32 mg/dL urea nitrogen (reference range, 15 to 44 mg/dL), and 59 U/L alkaline phosphatase (reference range, 40 to 126 U/L). Serum phosphorus was within normal limits (4.2 mg/dL; reference range, 2.5 to 4.5 mg/dL), as was vitamin D (65 ng/mL; reference range, 35 to 70 ng/mL).

Chest radiograph was unremarkable. Radiographs of both hips and shoulders showed multiple lobulated, calcified nodules with a periarticular distribution that seemed to involve the bursae (Figure 1). Distribution of lesions was confirmed with pelvic computed tomography scan (Figure 2). Calcifications were dense, nodular, and cystic; were confined to soft tissue; and extended mainly over the femoral extensor surface. Furthermore, one of the cystic lesions (Figure 2b) seemed to have



Figure 1. Massive tumoral calcinosis of shoulders and hips. Anteroposterior radiographs of (A) right shoulder, (B) left shoulder, and (C) both hip joints and (D) lateral radiograph of left femoral.

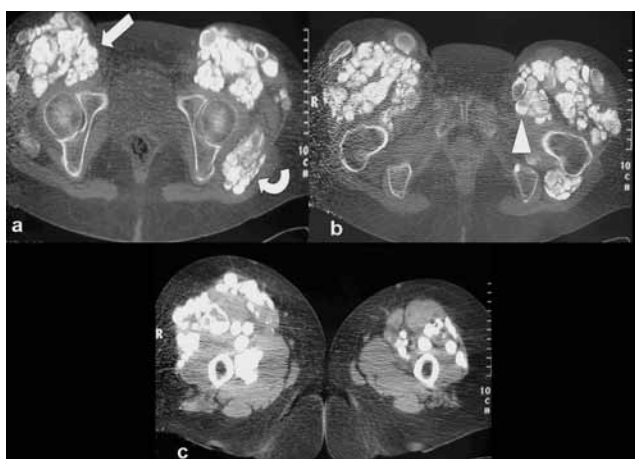


Figure 2. Computed tomography of tumoral calcinosis. (A) Extensive calcifications involve bursae bilaterally, mainly anterior part (arrows) and to lesser extent posterior (curved arrows). (B) Tumoral calcinosis appears as nodular and cystic calcifications, one of which shows sedimentation (arrowhead). (C) Calcifications extend to extensor surface of femoral region.

fluid-fluid levels caused by calcium layering (sedimentation sign). Left femoral calcifications involved, to a lesser extent, the flexor surface (Figure 2c). Although this massive calcinosis involved the anatomical region of the bursae, it was not confined to them but extended to soft tissues of the femoral extensor surface.

DISCUSSION

Tumoral calcinosis is a rare clinical entity initially described with the term *endotheliome calcifie* in 1898 by Giard¹ and in 1899 by Duret.² In the European literature, it became known as *Teutschlaender disease*, as it was Teutschlaender^{3,4} who studied this disease from 1930 to 1950. In 1943, Inclan and colleagues⁵ first used the term *tumoral calcinosis* to describe a specific disease manifesting with periarticular, calcified masses in the soft tissues with concomitant elevated serum phosphate but normal calcium levels. Inclan and colleagues specified that this condition occurred in the absence of renal, metabolic, or collagen vascular disease. Classic tumoral calcinosis lesions were characterized as lobular, densely calcified masses consisting of pleomorphic calcium phosphate (hydroxyapatite) crystals and confined to the soft tissue, generally at the extensor surface of the joint in the anatomical distribution of a bursa. The most common locations of tumoral calcinosis in descending order are hip, elbow, shoulder, foot, and wrist. Rare locations also include dental and paraspinal regions.⁶ As it was proved many years later,^{7,8} the condition Inclan and colleagues described is a rare hereditary disease. Although the pattern of inheritance is still debated, the generally accepted mode of transmission is autosomal dominant with variable penetrance.

Over the years, however, this term gradually came to incorporate any multilobulated calcification with periarticular distribution. In an attempt to rectify this

imprecision, Smack and colleagues⁹ classified tumoral calcinosis into 3 categories based on its pathogenesis: primary normophosphatemic tumoral calcinosis, primary hyperphosphatemic tumoral calcinosis, and secondary tumoral calcinosis.

Secondary tumoral calcinosis, which may be associated with renal failure, hemodialysis, collagen vascular disease (particularly scleroderma or dermatomyositis), sarcoidosis, pseudoxanthoma elasticum, massive osteolysis, and other conditions, is not as rare as primary forms.⁶

Pathogenesis of secondary tumoral calcinosis is based on chronic soft-tissue inflammation and is thought to represent a form of dystrophic calcification. It usually involves subcutaneous tissues, and it is circumscribed (*calcinosis circumscripta*). Calcinosis can occur in systemic sclerosis (*scleroderma*), but it is usually subcutaneous and limited in extent. Robertson and colleagues¹⁰ reported that calcinosis occurs in about 25% of patients with scleroderma, usually those with the disease limited to the skin, but not in tumoral form. It involves small joints (metacarpophalangeal, proximal, or distal interphalangeal joints) and is associated with anticentromere antibodies or CREST (calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, telangiectasia) syndrome. A tumor-like calcification of large joints is uncommon, occurring in less than 1% of patients with scleroderma.¹¹ To our knowledge, only 2 cases of scleroderma and tumoral calcinosis of large joints have been reported. The first case involved the left shoulder region. In that patient, the tumor-like calcifications were remarkably less extensive than our patient's, as they involved only a single shoulder region.¹² The second case involved a 67-year-old woman with calcific deposits within the shoulder region and larger deposits involving the fingers, elbow, wrist, and foot. That patient was diagnosed with Sjögren syndrome and systemic scleroderma.¹³

On the other hand, several cases of limited spine tumoral calcinosis related to scleroderma have been reported.¹⁴⁻²⁰ All the spine tumoral calcinosis cases are less extensive than the case presented.

CONCLUSION

Tumoral calcinosis is a rare clinical entity that can be a primary condition or a condition secondary to disease, such as renal failure, hemodialysis, sarcoidosis, dermatomyositis, or systemic sclerosis. In our patient, who had systemic sclerosis, development of calcifications was unique, as they involved many major joints and their extent was remarkable.

AUTHORS' DISCLOSURE STATEMENT

The authors report no actual or potential conflict of interest in relation to this article.

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