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Calcinosis Cutis Complicated with Recurrent Tissue Infections in a Patient with Limited Cutaneous Systemic Sclerosis: A Case Report

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ABSTRACT

Calcinosis related to systemic sclerosis is a debilitating vascular complication that negatively affects quality of life. It results from the deposition of calcium hydroxyapatite in soft tissues that occurs in the presence of normal calcium and phosphorus metabolism. Injuries can be painful and cause tissue inflammation, ulcers with infections, or deformities that cause functional disability.

We present the case of a 29-year-old woman with a history of calcinosis cutis associated with systemic sclerosis, lesions complicated by soft tissue infections, leading to septic arthritis. During the approach, improvement was achieved with the use of intravenous antibiotics and a long oral regimen. As it is a disease without definitive treatment, we improve the patient's living conditions and prevent future infections with non-pharmacological therapeutic measures.

KEYWORDS: calcinosis cutis, scleroderma, systemic sclerosis, cellulitis, septic arthritis.

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INTRODUCTION

Soft tissue calcification is divided into 5 subtypes: dystrophic, metastatic, idiopathic, tumor, and calciphylaxis. Systemic autoimmune rheumatic diseases are associated with the dystrophic type, characterized by the deposition of calcified material in damaged tissue with normal serum calcium and phosphate levels. [1]

The prevalence of calcinosis ranges from 18% to 49% in systemic sclerosis and appears similarly in both limited and diffuse cutaneous sclerosis, often occurring more than 10 years after diagnosis. Calcinosis most commonly affects areas of local trauma, such as areas exposed to repetitive pressure and friction, including the extensor areas of the elbows, fingertips, and thumbs. The condition is more common in men and is highly associated with other vascular complications, such as digital ulcers, acro-osteolysis, telangiectasias, and pulmonary hypertension. [2]

The pathophysiology behind dystrophic calcification is unclear. Calcinosis in systemic sclerosis is associated with vascular dysfunction and defective angiogenesis. These generate hypoxia in the tissues and increase the products of oxidative stress, interleukin (IL)-1, IL-6, IL-1b, and tumor necrosis factor, thereby causing tissue necrosis and releasing denatured proteins that promote calcification. Mitochondria can accumulate large amounts of calcium and phosphorus under prolonged inflammatory conditions and can be released during muscle damage, thus acting as nucleation sites for calcinosis. Lesions may be asymptomatic or painful. There may also be soft tissue inflammation, ulcers with superimposed infections, or even deformities leading to functional disability. [3]

CLINICAL CASE

A 29-year-old Caucasian female with a history of calcinosis cutis presented with a soft tissue infection in the right thigh. She had a history of systemic sclerosis and calcinosis cutis at the age of 15, along with sclerodactyly of the fingers, telangiectasia, salt and pepper lesions, occasional Raynaud's phenomenon, and anti-Scl70 antibodies. She was being treated with mycophenolate mofetil 500 mg and warfarin 2.5 mg, once a day. The patient also had a history of multiple calcium resection surgeries in her elbows and was hospitalized 5 months prior for septic arthritis in the right knee as a complication of an infected calcium lesion. The lesion required surgical lavage and intravenous antibiotic therapy because of streptococcus dysgalactiae. She

subsequently developed grade-4 knee gonarthrosis, which had begun 15 days prior with the release of a mousse-like material from a lesion on the right thigh (Figure 1). This was followed by erythema with the release of a purulent material that caused pain, edema, and fever (Figure 2). Upon admission, she showed normal vital signs with no evidence of shock. A physical examination of the right thigh showed evidence of an infection due to an injury with discharge of purulent material, calcium lesions in the knee, and limitation in range of mobility. We did not determine vascular compromise of the extremity. Meeting criteria for sepsis without severity data (leukocytosis 17,000/mm3, temperature >38°C, heart rate >90). We requested imaging of the right knee. An X-ray showed calcinosis in the soft tissues and grade-4 gonarthrosis. USG did not determine joint involvement. It was limited only to the soft tissues.

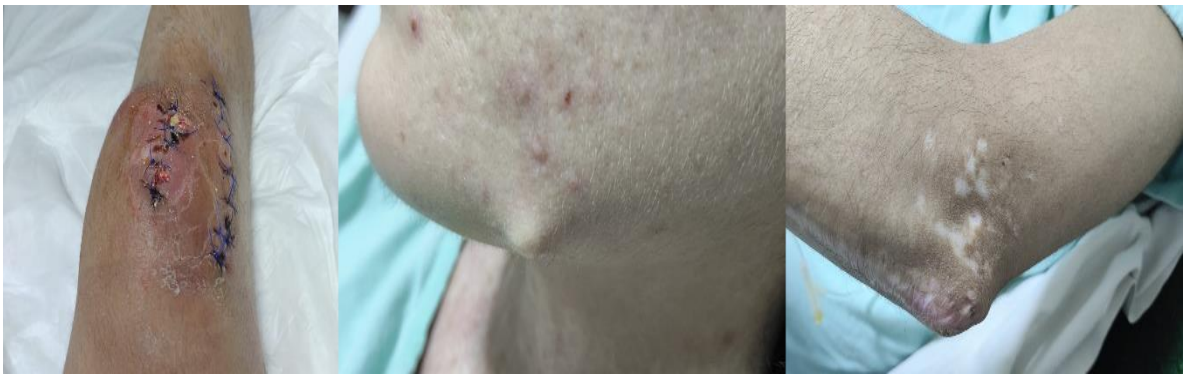

Figure 1. Calcinotic lesions in the jaw and elbows. Right knee with a history of septic arthritis.

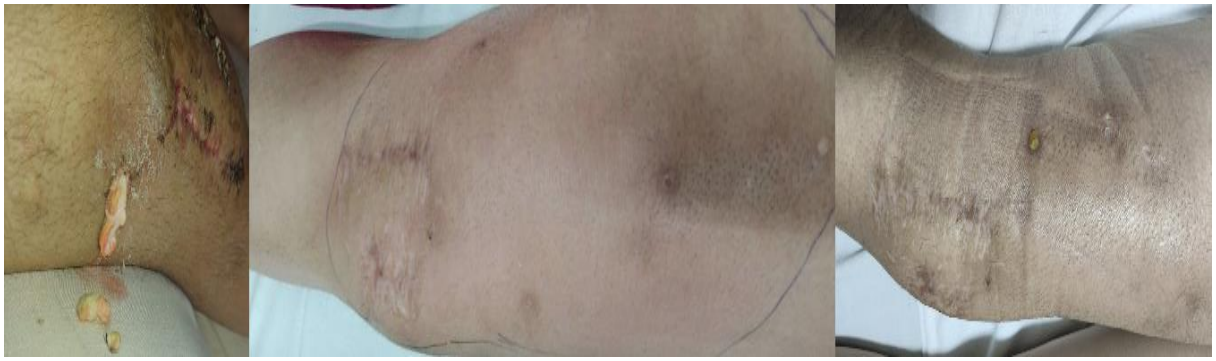

Figure 2. Clinical evolution of tissue infection in the right knee.

We adopted a multidisciplinary approach with rheumatology, infectology, and traumatology services. The simple tomography of the pelvic limb showed deep calcium lesions in the thigh, forming an exoskeleton, and confirmed no joint involvement, so we did not consider surgical treatment (Figure 3). We suspended treatment of the rheumatological disease during the acute infectious condition. Because data on hemodynamic instability were not presented and wound secretion culture had little efficacy in determining the agent, we considered monitoring the response to empirical treatment. Because of a history of previous infections and hospitalization, we began antibiotic therapy with intravenous doxycycline 100 mg every 12 days because of the risk of Methicillin-resistant Staphylococcus aureus. The patient showed an improvement in the infection and a decrease in the systemic inflammatory response with empirical antibiotic therapy before 72 hours, so we decided to discharge her from the hospital with a long regimen of 14 days at the same oral dose (Figure 2). During the follow-up in the outpatient clinic, the infection was completely resolved. We restarted treatment with mycophenolate mofetil and analgesics. We decided not to continue warfarin for the calcinosis cutis because no improvement was demonstrated during its use. Finally, we explained to the patient the importance of adequate hygiene for injuries and scheduled a follow-up visit.

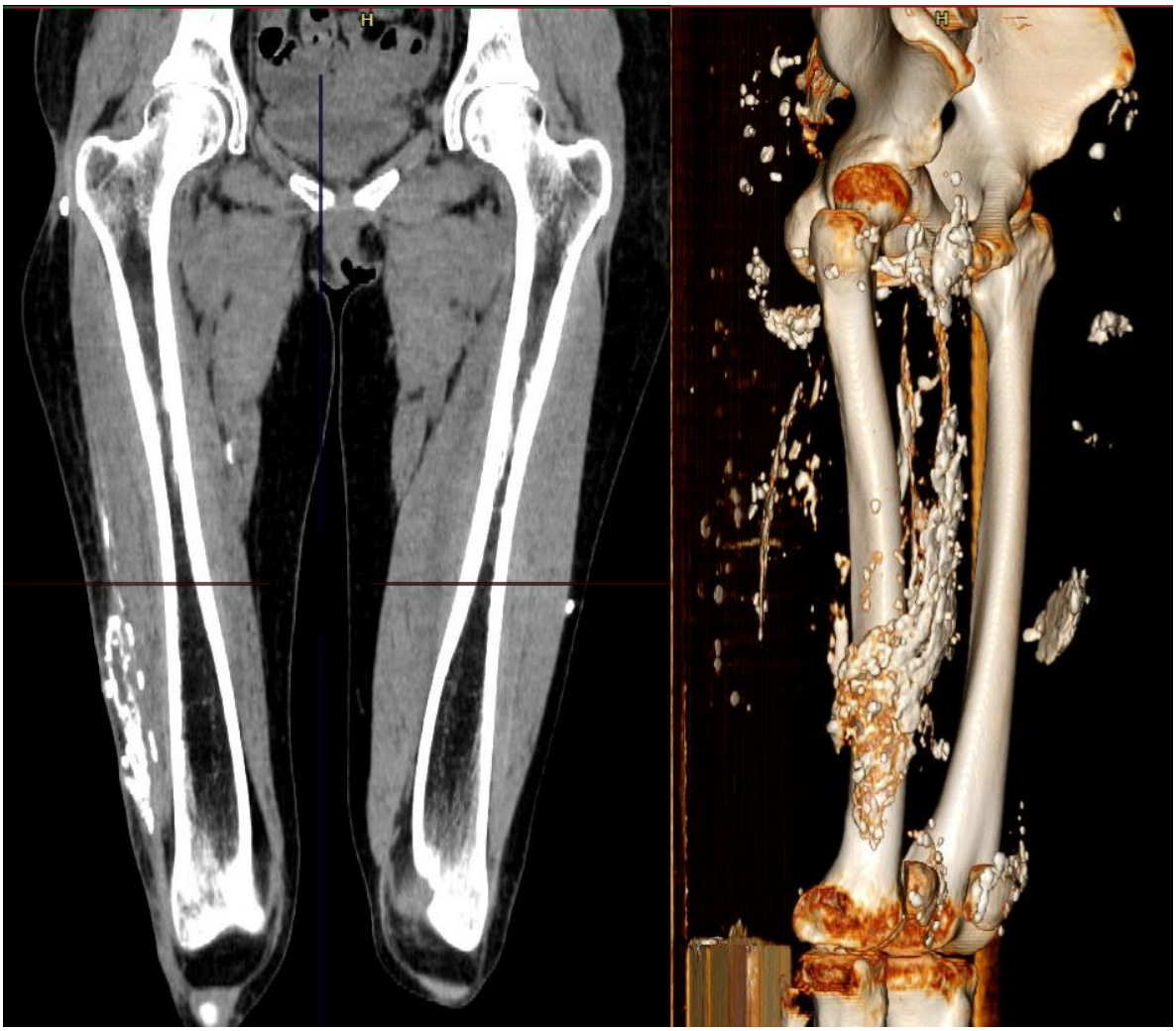

Figure 3. CT scan showing calcifications in the subcutaneous tissue and 3D reconstruction showing an exoskeleton due to the calcinotic lesions.

DISCUSION

Calcinosis cutis lesions, such as subcutaneous nodules, are the most common form in patients with systemic sclerosis. They affect the skin, subcutaneous fat, muscles, and tendons and may occur along myofascial planes or create extensive deposits that cover larger surface areas, forming an exoskeleton. They occur most frequently in the hands (65%–83%), proximal upper extremity (27%), knee or proximal lower extremity (10%–22%), and hip (6.7%). Injuries can cause pain or compression neuropathies with motor and sensory deficits; soft tissue edema; ulcers with infection; and deformities that cause functional limitations, mainly in the hands and elbows. [1-2]

Anticentromere and anti-PM/Scl antibodies have long been associated with a higher prevalence of calcinosis in patients with cutaneous systemic sclerosis. Cutaneous systemic sclerosis and anti-topoisomerase antibody (Scl-70) were predictors of calcinosis in a cohort of 1,305 patients from the Canadian Scleroderma Research Group registry, which includes patients from Canada and Mexico. Antinucleolar and anti-Scl-70 antibodies were more prevalent in Mexican patients with calcinosis. [4-5]

Plain radiography and ultrasound are sensitive for detecting calcinosis and are the first-line imaging modality. Computed tomography can better evaluate the extent of calcinotic lesions and provides information about surrounding structures such as neurovascular bundles. Magnetic resonance imaging provides better visualization of edema or inflammation. Depending on the shape and consistency, lesions can be classified as mousse (soft with a toothpaste-like liquid), network (thin and diffuse), plaque (large and uniform), or stone (single or multiple hard stones). The resolution time varies by form: network (22–140 days) and calculations (22–140 days). Clinical categorization is important for treatment and prognosis. [3-4]

There is no cure for calcinosis, and it remains a therapeutic challenge in patients with rheumatic diseases. General medical recommendations for the treatment of calcinosis include avoiding trauma and improving blood flow to the extremities. Treatment of Raynaud's phenomenon and digital ulcers may also have a preventive role in calcinosis. A multidisciplinary approach involving rheumatology, dermatology, and plastic surgery is recommended. Calcinosis lesions associated with cutaneous systemic sclerosis rarely

improve; most remain stable or worsen. Topical therapies, such as neem oil and Hypericum perforatum, can soften and facilitate the removal of the calcinotic material. In addition, topical or intralesional sodium thiosulfate (1 ml/cm^2) can act as a calcium chelator. Nonsteroidal anti-inflammatory agents and opioids may also be used to relieve pain. [1,6]

Pharmacological treatment is based on the use of less expensive medications with broad efficacy. Diltiazem, 60 mg 3 times daily, reduces intracellular calcium entry into affected tissues by influencing intracellular calcium levels in macrophages. Oral colchicine at a dose of 1 mg/day alters chemotaxis and phagocytosis of leukocytes by inhibiting microtubule polymerization, thereby reducing inflammation. Warfarin, a vitamin K antagonist, reduces MGP levels by preventing the carboxylation of glutamic acid. Minocycline, in doses of 50 to 100 mg per day for 4–8 months, chelates calcium and inhibits collagenolytic enzymes, reducing inflammation and ulceration. Corticosteroids, as a first-line treatment, act quickly to stop the disease process. The most reported treatment regimen was oral prednisolone or prednisone at a dose of 2 mg/kg/day and pulses of IV methylprednisolone (MP) at 30 mg/kg/day, followed by oral prednisone. [1,3]

Non-pharmacological interventions are typically reserved for injuries that cause chronic pain or compression neuropathy. Surgical excision is indicated in large, localized, and symptomatic lesions located on tendons, blood vessels, and nerves. Excision is not considered an optimal therapeutic option because of the increased risk of slow wound healing, infection, and a possible decreased range of motion. An alternative option is to use a high-speed dental bur instead of a scalpel, leading to better results, faster healing, and improved pain and function. The carbon dioxide laser is another option that is precise and causes less damage to surrounding healthy tissues. Extracorporeal shock wave lithotripsy uses acoustic waves to break up mineral deposits, has a high success rate, and is used in calcific tendonitis. [1,3]

Pain and erythema may be signs of soft tissue infection. Solid calcinosis may liquefy and ooze from the skin, leading to soft tissue infection. Drainage and a bacterial culture are recommended to evaluate antibiotic sensitivity. Topical antibiotics with lidocaine can treat minor soft tissue infections and relieve pain. Broad-spectrum oral antibiotics are used in moderate infections, whereas intravenous antibiotics are required for progressive soft tissue infection. Some patients are susceptible to recurrent cellulitis infections. A previous episode of cellulitis has a recurrence rate ranging between 8% and 20% annually, especially if it occurs in the legs. Staphylococcus aureus is the most common isolate from acute purulent soft tissue infections. The recommended duration of treatment is 5 days; if this period does not improve, the treatment should be prolonged. MRSA coverage is not recommended for nonpurulent cellulitis unless patients have failed initial antibiotic treatment or exhibit systemic inflammatory response syndrome and hypotension, cellulitis associated with penetrating trauma, or evidence of MRSA infection elsewhere. Oral antibiotics with MRSA coverage may be chosen, such as trimethoprim-sulfamethoxazole, clindamycin, doxycycline (100 mg orally twice daily), and linezolid. Furthermore, systemic signs of infection warrant the use of intravenous antibiotics. However, the benefit of intravenous versus oral administration of antibiotics for cellulitis of a similar severity has not been shown to be significant. [1,7-8]

CONCLUSION

Calcinosis is a common disease in patients with systemic sclerosis, associated with a longer time to diagnosis. There are different morphologies of calcium lesions, which produce chronic pain leading to functional limitations. Even though this disease is a therapeutic challenge, prevention and timely treatment can help prevent the formation of lesions, improve the patient's living conditions, and avoid complications such as soft tissue infection. Our patient had multiple injuries that predominated in the pelvic limbs, forming an exoskeleton. This area is associated with a greater risk of infections. Last, non-pharmacological treatment and good hygiene of the lesions can prevent the need for antibiotic therapy, except in the case of a local or systemic infection, always taking into account the risk of MRSA.

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