

Dystrophic calcinosis: do we really know how to treat it?

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INTRODUCTION

Calcinosis cutis is the deposition of calcium in the skin and subcutaneous tissue. It's a rare disorder occurring more frequently in systemic sclerosis and dermatomyositis¹⁻⁵.

The classification of soft-tissue calcifications includes the metastatic, tumoral, dystrophic and idiopathic types. The most common is the dystrophic type, characterized by normal calcium metabolism (normal serum level of calcium and phosphate)^{1,3}.

It's a long-term manifestation and clinical features associated to it are digital ulcers, osteoporosis, anti-centromere antibody and anti-PM-Scl antibody^{1,2}.

We hereby present a case of a patient with treatment-refractory extensive calcinosis.

CASE REPORT

A 63-year-old female was diagnosed with CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telangiectasias) 19 years ago. She currently reports inability to walk and dyspnea. On the clinical examination, she had masklike face, skin thickening and hardening (modified Rodnan score 26); major muscular atrophy and extensive calcinosis on shoulders, elbows, thorax, abdomen, knees and feet with joint involvement, causing pain and recurrent minor infections, requiring antibiotics and surgery. On the laboratory results, she had normocytic and normochromic anemia, Erythrocyte Sedimentation Rate (ESR) = 110mm, calcium = 8.3mg/dl, phosphorus = 4.1mg/dl, alkaline phosphatase = 131U/l and Creatine Phosphokinase (CPK) = 47U/l, negative anti-Scl-70 antibody, anticentromere antibody +, Antinuclear

Antibodies (ANA) = 1/240 centromere pattern. Chest, shoulders and hips x-rays (Figures 1A and B, 2 and 3) showed universal calcinosis.

During those 19 years, in addition to treatment for systemic sclerosis (nifedipine, D - penicillamine, prednisone and azathioprine), different calcinosis treatments were used, including diltiazem, colchicine, bisphosphonates (pamidronate and zoledronic acid), warfarin and surgery, but all were ineffective.

DISCUSSION

Calcinosis pathophysiology is not clear, but there are some theories that inflammation and vascular ischemia play a role in its process^{1,2,4}.

Diagnosis can be made by plain radiography, as it has a very good sensitivity to detect calcinosis and is indicated as initial imaging evaluation^{1,2,4}.

Management includes general measures, pharmacological and non-pharmacological treatment. As general measures, it is important to improve blood flow in extremities¹.

Pharmacological treatment includes diltiazem, which reduces intracellular calcium influx and alters calcium nidus formation. It's considered as a first-line approach. Warfarin acts at vitamin K levels, which is involved in the calcium-binding process, but studies results are contradictory^{1,2,4}.

Biphosphonates may be used to inhibit osteitis and bone resorption and have shown positive results. Colchicine is not effective in reducing calcium deposit sizes, but works on reducing local inflammation. Patients treated with rituximab seem to have beneficial results, but more studies are required to infer something concrete¹⁻⁴.

Finally, surgery is indicated mainly in localized calcinosis or when there is major pain or function loss²⁻⁴.

As observed, our patient had the main treatment options for calcinosis and they were not effective. Cur-

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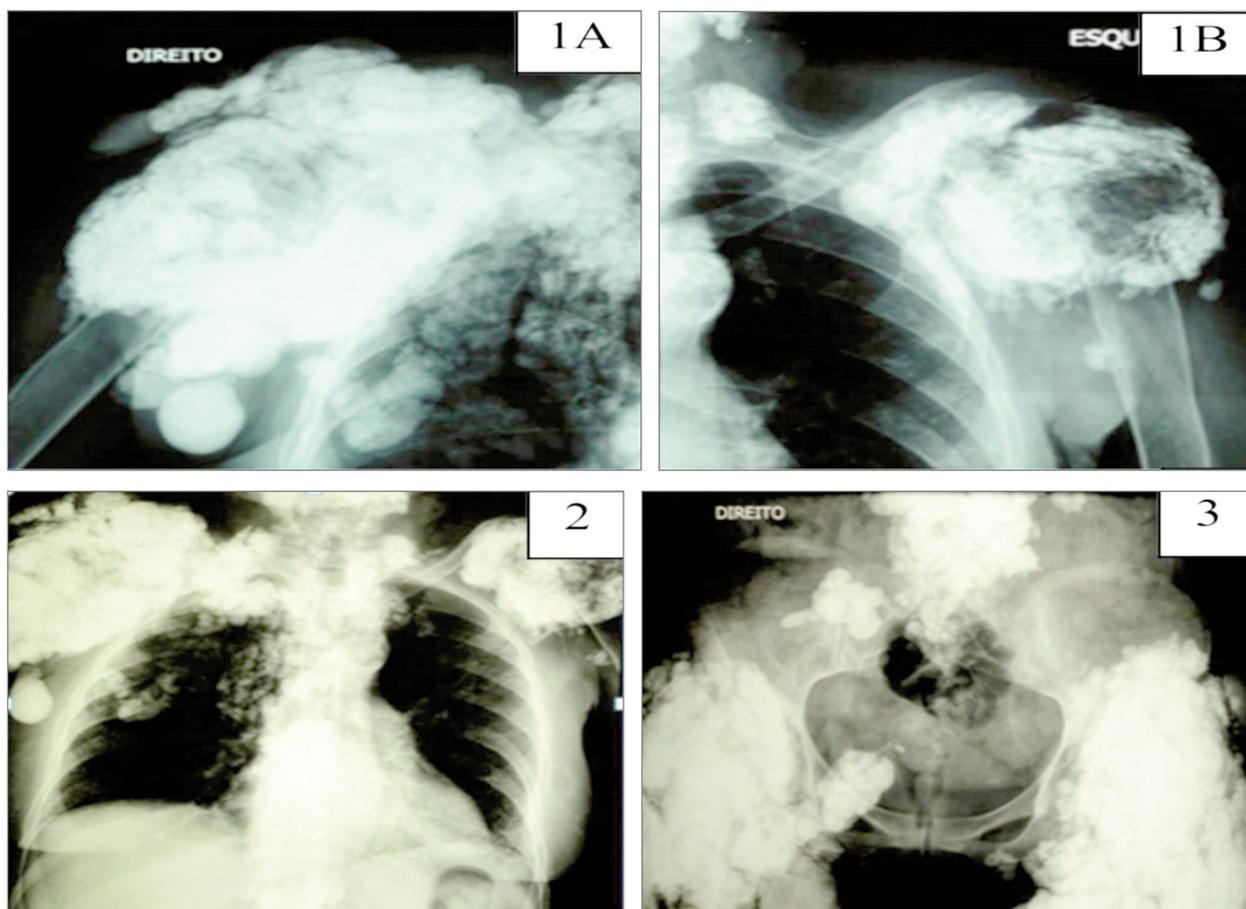


FIGURE 1A and B, 2 and 3. Extensive calcinosis lesions – shoulders, thorax and hips

rently, she still lives with a lot of pain and extensive calcinosis all over her body. In light of the above, we wonder, do we really know how to treat calcinosis? More studies are required for more robust conclusions and to improve our patients' quality of life.

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