# Treatment of cutaneous calcinosis in CREST syndrome by extracorporeal shock wave lithotripsy

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We describe the unusual case of a 78-year-old woman consulting for extensive and painful wound leg ulcerations and calcifications secondary to CREST syndrome that was treated by extracorporeal shock wave lithotripsy. This treatment was considered because of the severity of our patient's symptoms and her failure to respond to various medical and surgical treatment. (J Am Acad Dermatol 2005;53:S263-5.)

CREST syndrome is a variant of systemic sclerosis, which affects a relatively small number of patients. CREST (also called limited cutaneous scleroderma) has a more favorable prognosis than the diffuse form of scleroderma. However, cutaneous calcinosis, which occurs during the disease course, is always difficult to manage. There are very few satisfactory medical and surgical therapies for cutaneous calcinosis in connective tissue disorders. For these reasons, we tried a new therapeutic approach, extracorporeal shock wave lithotripsy (ESWL). ESWL, a minimally invasive treatment, plays a major role in the treatment of urolithiasis. It has a high success rate, and the spectrum and incidence of treatment-related morbidity are low. This technique has not been previously reported for the treatment of cutaneous calcinosis in scleroderma.

## CASE REPORT

A 78-year-old woman developed painful, large ($\leq$20 $\times$ 12 cm) ulcers with calcinosis on her lower limbs. She described similar lesions earlier in 1991 and 1999 that healed after skin grafting. In 2001, a limited systemic sclerosis of the CREST type (calcinosis, Raynaud's syndrome, esophagal mobility disorders, sclerodactyly, and telangiectasia) was diagnosed with a peptidic esophageal stenosis that was then dilated. High titers of antinuclear antibodies with specificity for centrosomes (1:10240) were detected. Routine laboratory tests revealed anemia and hypoalbuminemia. The patient was normocalcemic and normophosphatemic. Lung diffusion capacity for carbon monoxide, cardiac function, and renal function were all normal.

Mechanical debriding was limited as a result of significant pain despite local anesthesia (Emla cream 2 hours prior).

Because of the distribution of the ulcers over her left and right legs, which extended to the back of her legs, the patient was referred to operation. Angiography showed normal vessels in both legs. She was treated with partial excision of calcifications and skin grafting under general anesthesia.

A few months later, after initial improvement, leg ulcers remained refractory to conservative treatment, and numerous ulcers reappeared. They again required skin grafting. Low-dose minocycline was introduced but neither wound healing nor disappearance of the calcifications were observed.

Two years later, the patient was found to have unusually severe calcinosis cutis in both legs extending from both knees to ankles. ESWL was suggested to destroy dystrophic calcifications to allow ulcers to heal. Clinical and radiologic follow-up of the calcinosis deposits was performed and the patient's assessment of the degree of discomfort, size, and frequency of ulceration was recorded. ESWL resulted in precise ablation of superficial dystrophic calcifications 15 days after the session (Fig 1). The patient

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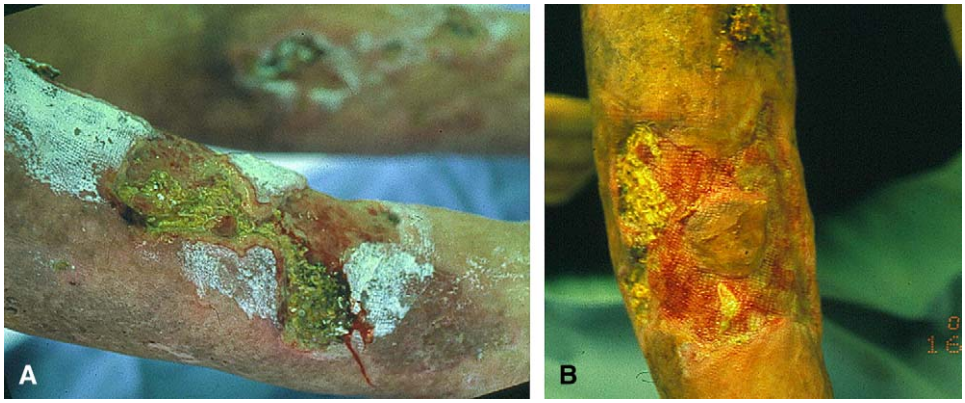


**Fig 1. A**, Large ulcer of leg with subcutaneous calcifications before first session of extracorporeal shock wave lithotripsy. **B**, Stone-free and healing ulcer after second session.

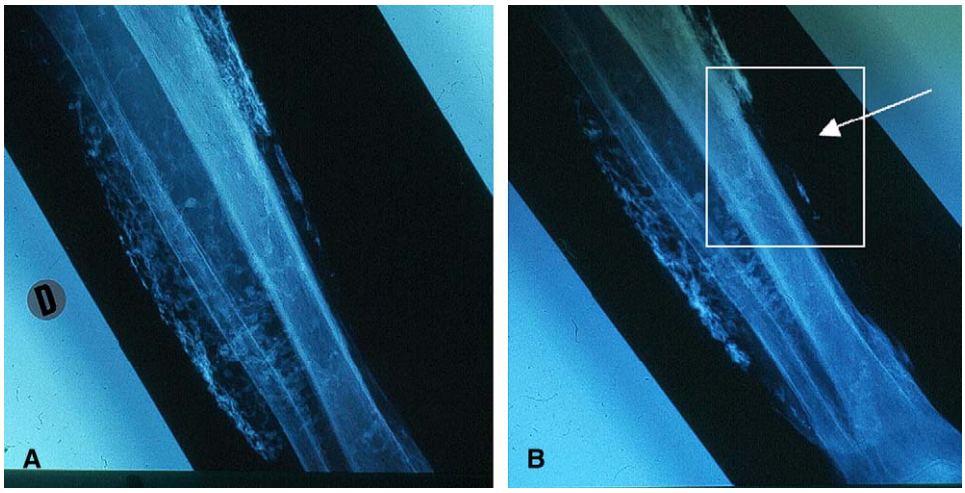


**Fig 2. A**, Extensive calcification on limb before extracorporeal shock wave lithotripsy. **B**, Disappearance of dystrophic calcification after one session of lithotripsy.

was stone free on radiography 1 month after the first treatment (Fig 2). The treatment was well tolerated and the size of ulcers and pain diminished after the first session. No immediate side effects from the lithotripsy were noted. Two others sessions were performed after 2 months. In the meantime, we noted decreased microcalcifications, and the ulcers started to heal (12 × 7 cm after 3 sessions). Unfortunately, the patient died after femoral fracture.

## DISCUSSION

Subcutaneous calcifications often appear in connective tissue disorders and are difficult to manage. Dystrophic calcifications are responsible for pain and infirmity when calcinoses are located close to articulations. Calcification of soft tissue may be a nonspecific local response or a symptom of complex underlying disease. Patient approach and treatment may vary depending mainly on the origin of soft tissue calcifications. To date, the physiopathology remains unclear.

Therefore, two therapeutic approaches can be proposed. On one hand it is possible to stop the progression of the dystrophic calcifications and on the other hand to destroy calcinosis to reduce pain and allow skin healing.

Unfortunately the medical treatment of calcinosis is uniformly disappointing.

Metzger et al[1] reported the failure of disodium etidronate to treat calcinosis in dermatomyositis and scleroderma. Edathamil has been used to treat calcinosis with success.[2] Vayssairat et al[3] showed that diltiazem does not reduce subcutaneous calcinosis. Hazen et al[4] noted the efficacy of intralesional

adrenal steroids in calcinosis in a patient with scleroderma. Robertson et al[5] reported the efficacy of low-dose minocycline to control calcinosis in systemic sclerosis in 9 patients with limited systemic sclerosis, reducing the size of calcinosis deposits and decreasing the frequency of ulceration and inflammation associated with calcinosis. The mechanism of action is probably mediated through the inhibition of matrix metalloproteinases and anti-inflammatory effects.

A low-dose warfarin regimen (1 mg/d) appears to have no demonstrable adverse effects. Previously published results suggest a beneficial effect on the progression of calcinosis in calcinosis universalis secondary to dermatomyositis or systemic sclerosis. Cukierman et al[6] evaluated the effect of low doses of warfarin in 3 patients with systemic sclerosis with disseminated subcutaneous calcinosis for 1 year. Two of the patients, with relatively small calcinotic lesions, responded to warfarin treatment, with complete resolution of the calcinosis. The other patient, with larger and older lesions, did not respond to treatment. Warfarin appears to be a good alternative to treat small and recent calcifications.[6] Yoshida et al[7] also described the use of low-dose warfarin to treat calcinosis in an ulceration of the finger in a patient with systemic sclerosis or CREST syndrome. After beginning warfarin, no calcium-containing substance was discharged from the fingertip.

On the other hand, the destruction of dystrophic calcifications remains necessary to limit painful and extensive ulcerations. Mechanical debriding of large calcinoses is often difficult despite local anesthesia. Surgical therapy requires skin grafting.[8] Chamberlain and Walker,[9] using carbon-dioxide laser vaporization, observed a remission of symptoms in cutaneous calcinosis linked to CREST syndrome.

The case of our patient with CREST syndrome—associated calcinosis suggest that ESWL is an effective treatment. Unfortunately the follow-up period was shortened by the premature death of the patient caused by an unrelated event. This is the first case of successful ESWL calcinosis treatment in CREST syndrome to our knowledge. Recently, Chan and Li[10] reported the case of a 23-year-old Chinese man with dermatomyositis-associated calcinosis cutis treated with ESWL. They observed a reduction in pain after two courses of ESWL.

We conclude that lithotripsy may offer effective remission of symptoms in cutaneous calcinosis in CREST syndrome and healing of ulcerations. Lithotripsy facilitates partial removal of calcium deposits and improves the size of ulceration within a few treatments. ESWL is a safe and well-tolerated procedure that can be proposed to patients with subcutaneous calcinosis.

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