

# Extensive skin necrosis of the arm in a patient with complex regional pain syndrome

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## Summary

We report a 36-year-old woman with complex regional pain syndrome (CRPS) type 1 presenting with extensive skin necrosis of the left arm. The patient cooled her arm with ice packs to ease severe pain due to CRPS, in spite of repeated cautions against frostbite injury. The regions of skin necrosis corresponded with the sites where she had applied ice packs. We considered that the severe skin necrosis in our case was due to a self-induced frostbite injury.

Complex regional pain syndrome (CRPS) consists of clinical symptoms such as severe pain, autonomic dysfunction, trophic changes of the hair, nails and skin, and functional impairment such as weakness, tremor and dystonia.<sup>1–3</sup> The cutaneous features of CRPS include oedema, erythema, papules, atrophy, ulceration, pigmentation and bullae. Previous reports have suggested that some ulcerations occurring in association with CRPS might be factitial.<sup>4</sup> CRPS has been subdivided into type 1 where there is no causative nerve damage, and type 2 where there is damage. We describe a patient with CRPS type 1, who developed widespread skin necrosis on her left arm as a result of excessive cooling with ice packs.

## Report

A 36-year-old woman presented with an intractable small ulcer, 5 mm in diameter, on the dorsal side of the left index finger, and persistent pain in the entire left forearm. Two months before her visit, she had noticed a small red area on the left index finger. At that time, a skin biopsy had been taken from the erythematous area

because it was refractory to topical corticosteroid ointment and systemic antibiotics. Histopathological examination found nonspecific perivascular inflammation in the dermis. After the biopsy, the erythema on the finger ulcerated with severe pain and swelling, which spread to the entire left forearm.

On physical examination, there was marked swelling, redness and warmth over the entire left arm. An oedematous, erythematous area with many small bullae was seen on the dorsal side of the left wrist (Fig. 1a). The patient reported that she cooled her left arm with ice packs all day to alleviate pain.

Results of blood tests including C-reactive protein and white blood cell count were unremarkable. Tests for both antinuclear antibody and rheumatoid factor were negative. Neurological examinations revealed hyperalgesia and sensory testing-induced paraesthesia in the left arm, and global weakness of all of the muscles of the left hand.

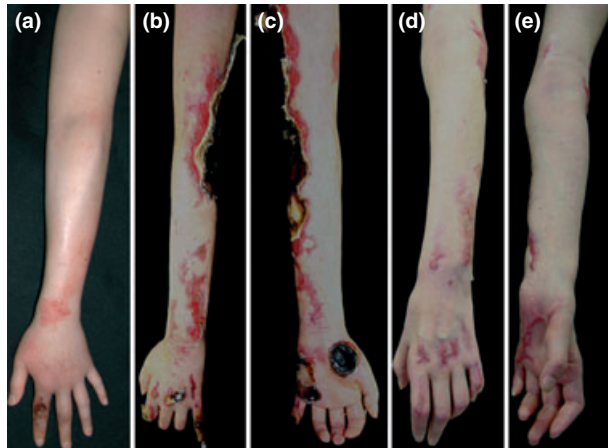
After admission to our hospital, a further biopsy specimen was taken from the left index finger, which revealed nonspecific granulation tissue. Cultures for bacteria were negative, as was PCR for mycobacteria. Thermography showed that the temperature in the left arm was 1 °C higher than in the right. Repeated psychiatric assessment was unremarkable.

Based on these data, and according to the diagnostic criteria,<sup>1</sup> we diagnosed the skin lesions as CRPS type I. Treatment with non-steroidal anti-inflammatory drugs, oral steroids, triptanol, depromel and opioids for the

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**Figure 1** (a) Initial examination showing oedema, erythema and small bullae on the dorsal aspect of the wrist; (b, c) extensive skin necrosis around the elbow and hand on readmission (4 months after the first admission); (d, e) 18 months after the onset of CRPS, severe muscle atrophy and contraction of the elbow, wrist and fingers were observed.

severe pain had no effect. Sympathetic nerve block was not performed because systemic phentolamine administration did not relieve the pain.

The patient voluntarily moved to another hospital in her home town, and despite repeated cautions against cold-induced injury, continued all-day cooling of the arm. Six months later, she was admitted to our hospital again with extensive skin necrosis on the left arm and a contracted left elbow joint (Fig. 1b,c). The areas of skin necrosis exactly matched the sites where she had applied ice packs.

Debridement was performed, after which the elbow was covered with a latissimus dorsi flap, and other parts of the arm were treated with occlusive dressings. We used continuous infusion of fentanyl citrate for expected postoperative pain for 1 week, followed by oral morphine hydrochloride. The severe pain gradually subsided and morphine was discontinued 4 months after the operation. Eighteen months after the onset of CRPS, the patient's left arm, hand, and fingers showed severe contraction and muscle atrophy (Fig. 1 d,e).

CRPS presents with various skin abnormalities including oedema, erythema, papules, atrophy, ulceration, pigmentation and bullae.<sup>2,3</sup> Some ulcerations occurring in association with CRPS might be factitial, such as those reported in the paper by Lipp *et al.*,<sup>4</sup> who reported a case of CRPS with mutilating ulcerations suspicious of a factitial origin in a patient with personality disorder. Webster *et al.*<sup>5</sup> reported a case of reflex sympathetic dystrophy with linear erosions, which were improved only by occlusive dressing. In our patient, we felt the linear skin necrosis, which corresponded to sites of ice-pack application was due to excessive cooling. Does improper use of an ice pack result in frostbite by direct cold injury? Evidence to support this hypothesis includes a case report that a

patient with acute synovial arthritis had applied frozen ice packs continuously for more than a couple of hours for 9 days, which resulted in frostbite with local skin necrosis.<sup>6</sup>

It remains unknown why the patient cooled her arm until skin necrosis developed; however, sensory disturbances are common in CRPS.<sup>7,8</sup> Pain in CRPS is often exacerbated by cold, heat or contact. Usually, cold intolerance in CRPS is a troublesome symptom in the winter and is probably due to regional impairment of vasomotor regulation.<sup>7</sup> However, in some cases, patients with CRPS show hyposensitivity to cooling. Birklein *et al.*<sup>8</sup> tested thermal thresholds in patients with CRPS and found a significant increase in heat perception thresholds and a significant decrease in cold pain thresholds on the affected side. Based on these observations, the pain our patient had might have been not only hypersensitive to stimulation by warmth but also hyposensitive to cooling.

Treatments for CRPS aim to achieve the relief of pain and the maintenance and restitution of function.<sup>1</sup> Pathophysiologically orientated therapy includes steroids, sympathetic blocks and radical scavengers such as dimethyl sulfoxide. Symptomatic treatment of neuropathic pain in CRPS includes antidepressants, anti-epileptic drugs and opioids. Non-drug treatments such as physical therapy are also essential to improve and maintain function and mobility of the affected limb. We could not stop our patient from cooling her arm because she insisted that it was the only way to improve the pain, although we tried every conceivable means including a tricyclic antidepressant, a selective serotonin reuptake inhibitor and repeated psychological counselling. The earlier introduction of a more effective treatment might have reduced this patient's considerable morbidity.

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