

## A Case of Complex Regional Pain Syndrome: an Underdiagnosed Condition in Dermatology

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Sir,

Complex regional pain syndrome (CRPS), also known as reflex sympathetic dystrophy (RSD) or causalgia, is a progressive and painful disorder that usually develops after trauma affecting the limbs. Although this disease is known to present with various cutaneous symptoms, there is little dermatological literature detailing the skin changes involved. We describe here a 25-year-old female patient with persistent pain and sclerotic and oedematous skin lesions in the left hand, which later progressed to the other hand. We also briefly review the updated epidemiology, spread pattern and treatment from the recent neurological and anaesthesiological literature, with the aim of raising awareness of this potentially important and easily missed disease.

### CASE REPORT

A 25-year-old woman presented to our department because of persistent pain and sclerotic and oedematous skin lesions on her left hand. One year prior to her first visit, she had accidentally sprained her left wrist. The initial pain subsided quickly, but after a month, severe burning pain developed on her left thenar eminence with marked swelling of the distal forearm and the hand. The pain gradually intensified and spread to the entire left hand, accompanying numbness, severe cold intolerance and muscle weakness. She underwent physical therapy for several months at an orthopaedic clinic with no effect.

On examination, pale-coloured, oedematous and indurated skin with a shiny surface was observed over the patient's left dorsal hand and fingers, mimicking sclerodactyly (Fig. 1). The lateral 3 digits showed mild flexion contracture. The dorsal skin over the left fingers could not be pinched. Hair loss and hypohidrosis were also observed.

Routine laboratory test results were unremarkable. Neurological examinations revealed hyperalgesia, paresthesia, dysesthesia and muscle weakness in the left forearm and the hand. Results of nerve conduction studies were normal. Radiographic examination showed marked decalcification of her left lateral 3 fingers (Fig. 1). Thermography revealed a 2.6°C lower temperature in the left dorsal hand than in the right. Eosinophilic fasciitis was excluded because this case showed unilateral involvement. The absence of autoantibodies and presence of neurological symptoms were inconsistent with scleroderma. Using the history of trauma, symptoms and physical findings, CRPS I was diagnosed. Treatment with regular stellate ganglion blockade and infra-red therapy was initiated. The therapy proved to be effective in relieving pain and hyperalgesia and in ameliorating the range of motion, although the effect was not long-lasting. Despite the treatment being continued for approximately 10 months, pain and oedema have now spread contralaterally to involve the right hand.

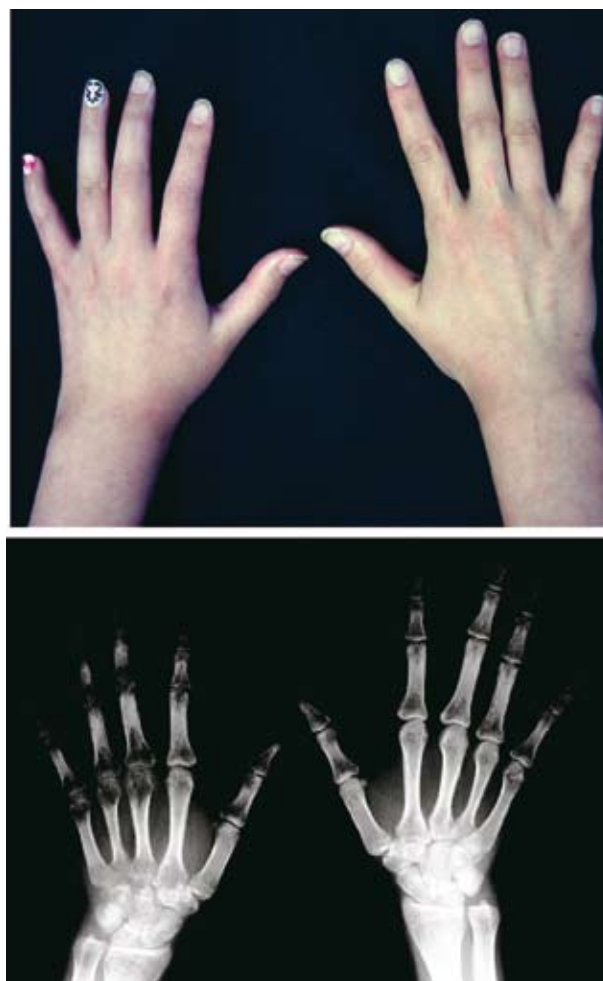


Fig. 1. Pale-coloured, oedematous and indurated skin with a shiny surface over the left dorsal hand and digits (upper panel). The lateral 3 fingers show mild flexion contracture. X-ray image showing marked decalcification of the left lateral 3 fingers (lower panel).

### DISCUSSION

CRPS/RSD has been reported to follow various injuries. According to the chart review of 134 patients by Allen et al. (1), the 4 most common inciting injuries are sprain/strain (29%), post-surgical (24%), fractures (16%) and contusion/crush injury (8%). No inciting event was remembered by 6% of patients. Other causes (11%) included venepuncture, laceration and spinal cord injuries. It has been proposed that immobilization plays a role in the pathogenesis of CRPS, because 47%

of cases had a history of physical immobilization with a cast or splint. Incidence of CRPS after a fracture is reported to be 1–2% (2). In the dermatological field, nail biopsy (3), nevus excision (4) and herpes zoster (5) have been reported as rare inciting events.

The diagnostic criteria for CRPS defined by the International Association for the Study of Pain are as follows: (i) preceding noxious event without (CRPS I) or with (CRPS II) obvious nerve lesion; (ii) spontaneous pain or hyperalgesia/hyperesthesia not limited to a single nerve territory and disproportionate to the inciting event; (iii) oedema, skin blood flow or sudomotor abnormalities, motor symptoms or trophic changes are present on the affected limb, in particular at distal sites; (iv) other diagnoses are excluded (6). The diagnosis of CRPS is primarily clinical and no specific test exists. However, several examination findings are helpful in supporting the diagnosis. Radiography typically shows spotty osteoporotic changes after 4–8 weeks in 40% of cases and thermography can reveal temperature difference of more than 1.0°C (6).

CRPS does not necessarily involve only one extremity. There are reports that it can spread from the initial site of presentation in various patterns. A retrospective analysis of 27 CRPS patients by Maleki et al. (7) revealed that all the patients experienced contiguous spread, which means a gradual and significant enlargement of the area affected initially. Unexpectedly, a large percentage of patients (70%) also experienced independent spread, e.g. first in a foot, then in a hand. Fifteen percent had mirror-image spread, e.g. first in the left hand, then in the right hand, as was seen in our patient.

The pathogenesis of CRPS has been emphasized to be due to excessive sympathetic adrenergic outflow. However, it has been speculated that failure of inhibitory spinal or supraspinal influences on nociceptive transmission might contribute CRPS development (8). The intrathecal administration of gamma-amino butyric acid (GABA)-receptor B agonist, which inhibits sensory input to the neurons of the spinal cord, successfully improved dystonia in CRPS patients (9). Furthermore, aberrant central nervous system regulation of neurogenic inflammation has also been suggested to play a role in both the initial presentation and the spread of CRPS.

Although block therapies have been traditional first-line treatment, there is little evidence to support their use (10). A stepwise functional restoration algorithm has recently been proposed in the consensus guidelines (11), in which physiotherapy including oedema control and desensitization are the mainstay of the treatment. Blocks, pharmacotherapy and psychotherapy are reserved for patients failing to progress to the next step in this algorithm. As no single curative treatment for CRPS exists, such a multidisciplinary approach is thought to be most helpful in pain control and restoration of function.

A rapid increase in the incidence of reported cases of CRPS occurred after the diagnostic criteria were published in 1994 (2). Nevertheless, general awareness of this condition seems still to be relatively poor, as observed in the study by Allen et al. (1) showing that an average of 30 months pass and 4.8 different physicians are seen before patients with CRPS are referred to a pain centre for adequate therapy. The condition does not seem to have gained much recognition among dermatologists, despite the fact that it manifests various cutaneous alterations (12). Out of 319 medical papers published in English in the past 5 years including CRPS or RSD in the title, only one report of ours (13) was found in the dermatological literature, presumably reflecting the scarcity of cases that are diagnosed in dermatological clinics. However, CRPS cases with cellulitis-like symptoms, Raynaud's phenomenon or painful oedema might be more likely to be referred to dermatologists at their first visit. Recognition and knowledge of this disease are essential, because delay in proper diagnosis and treatment may have a negative effect on patient outcome (1).

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