Complex Regional Pain Syndrome Type II After a Brachial Plexus and C6 Nerve Root Injury

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PRACTICE **POINTS**

- Complex regional pain syndrome (CRPS) is a neuropathic disorder of the extremities characterized by pain, a variety of autonomic and motor disturbances, and dermatologic findings.
- Early recognition of CRPS is critical, as it presents life-changing morbidities to patients.
- A multidisciplinary treatment approach with physical therapy, occupational therapy, psychological support, and pain control is needed for the management of CRPS.

To the Editor:

A 62-year-old man presented with an atrophied painful left arm of 17 years' duration that began when he was hit by a car as a pedestrian. He sustained severe multisystem injuries from the accident, including left brachial plexus and C6 nerve root avulsion injury. When he regained consciousness after 6 weeks in the intensive care unit, he immediately noted diffuse pain throughout the body, especially in the left arm. Since the accident, the patient continued to have diminished sensation to touch and temperature in the left arm. He also had burning, throbbing, and electrical pain in the left arm with light touch as well as spontaneously. He was thoroughly evaluated by a neurologist and was diagnosed with complex regional pain syndrome (CRPS) type II. For the treatment of pain, dorsal column stimulation and hemilaminectomy with exploration of the avulsed nerve root were attempted, both of which had minimal effect. He was maintained on hydromorphone, methadone, and oxazepam. He reported that for many years he was unable move out of bed due to the unbearable pain. With pain medications, he was able to regain most of his independence in his daily life, though the pain and other clinical aspects of CRPS still completely limited his use of the left arm.

Physical examination revealed glossy, cold, hairless skin with hypohidrosis of the left arm, forearm, and hand (Figures 1 and 2A). The left arm was conspicuously atrophied, with the forearm and hand erythematous. The fingers were taut, contracted, and edematous (Figure 2B), and the skin was unable to be pinched. The fingernails on the left hand had dystrophic changes including yellow color and brittleness with longitudinal ridges (Figure 3). The patient could activate the left bicep and tricep muscles against gravity but had minimal function of the deltoid muscle. He also had minimal movement of the left index finger and was unable to move any other digits of the left hand. The patient was continued on pain management treatments and physical therapy for his condition.

Complex regional pain syndrome is a neuropathic disorder of the extremities characterized by pain and a variety of autonomic and motor disturbances such as local edema, limited active range of motion, and vasomotor and trophic skin changes. There are 2 types of CRPS: type II is marked by explicit nerve injury and type I is not. The pathophysiology of CRPS is unknown.[1-3]

There is no definite set of diagnostic criteria for CRPS. The lack of any gold-standard diagnostic test for CRPS has made arriving at one valid, widely accepted set of diagnostic criteria impossible.[1] There are 4 widely used sets of diagnostic criteria. One is the International Association for the Study of Pain diagnostic criteria defined in 1994.[4] However, the criteria rely entirely on subjective symptoms and have been under great scrutiny

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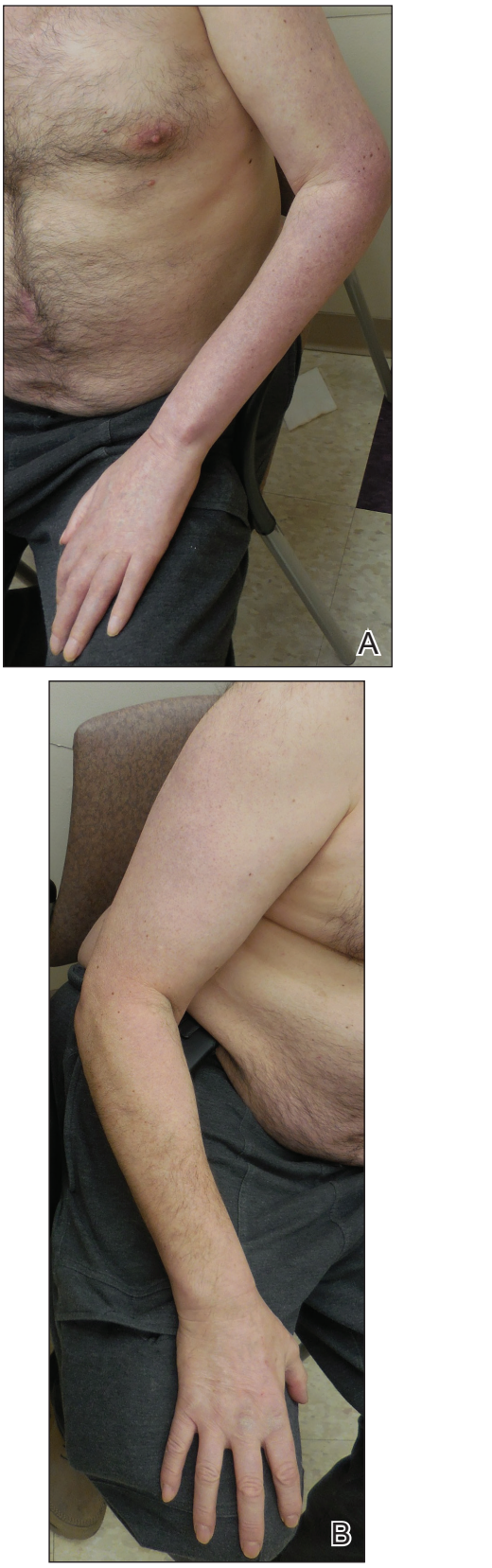

FIGURE 1. Complex regional pain syndrome type II. A, Glossy, cold, hairless skin of the left arm. B, The right arm was unaffected.

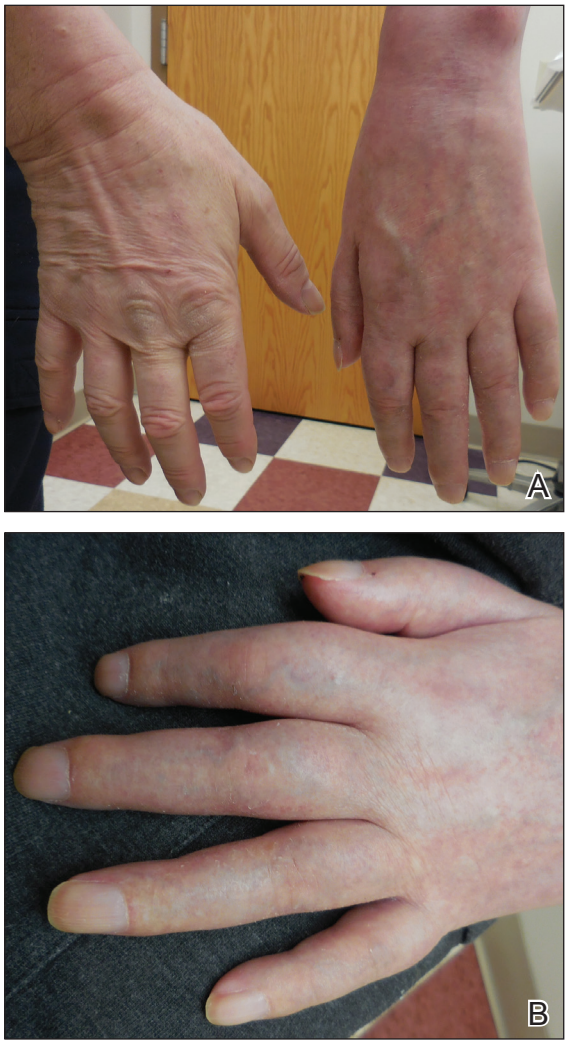

FIGURE 2. Complex regional pain syndrome type II. A, The left hand was erythematous. B, The fingers were taut, contracted, and edematous.

due to their questionable validity.[2] Veldman et al[5] presented other widely used CRPS diagnostic criteria in their prospective study of 829 reflex sympathetic dystrophy patients, which paid particular attention to the early clinical manifestations of CRPS. In 1999, Bruehl et al[2] proposed their own modified diagnostic criteria, which required physician-assessed signs in 2 of 4 categories to avoid the practice of exclusively relying on subjective symptoms. In addition, during a consensus meeting in Budapest, Hungary, a modified version of the Bruehl criteria was proposed.[6] All 4 criteria rely solely on detailed history and physical examination, and the choice of diagnostic criteria remains subjective.

The pathophysiology of CRPS also remains unclear. There are several proposed mechanisms such as sympathetic nervous system dysfunction, abnormal inflammatory response, and central nervous system involvement.[1] Psychologic factors, sequelae of nerve injury, and

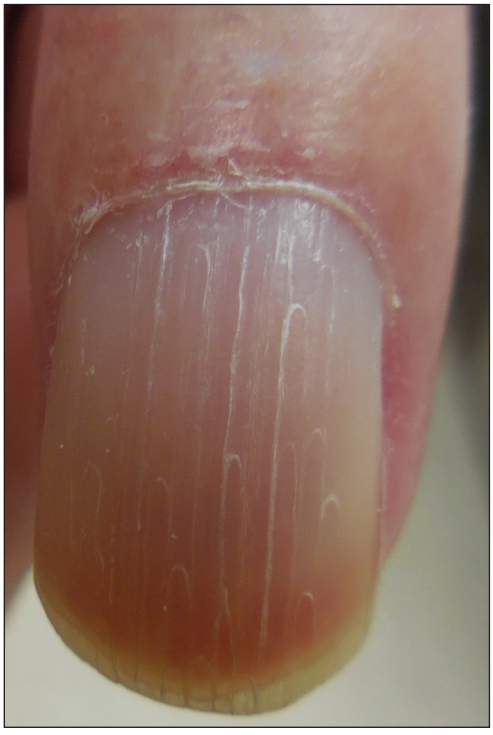

FIGURE 3. Complex regional pain syndrome type II. Yellow and brittle fingernail with longitudinal ridges.

genetic predisposition also have been implicated in the pathophysiology of CRPS.[1] It is likely that several mechanisms variably contribute to each presentation of CRPS.

Many dermatologic findings, in addition to neuromuscular symptoms, accompany CRPS and serve as important clues to making the clinical diagnosis. Complex regional pain syndrome has been thought to have 3 distinct sequential stages of CRPS.[1,3,7] Stage 1—the acute stage—is marked by hyperalgesia, allodynia, sudomotor disturbances, and prominent edema. Stage 2—the dystrophic stage—is characterized by more marked pain and sensory dysfunction, vasomotor dysfunction, development of motor dysfunction, soft tissue edema, skin and articular soft tissue thickening, and development of dystrophic nail changes. Stage 3—the atrophic stage—is marked by decreased pain and sensory disturbances, markedly increased motor dysfunction, waxy atrophic skin changes, progression of dystrophic nail changes, and skeletal cystic and subchondral erosions with diffuse osteoporosis.[1,3,7]

The staging model, however, has been called into question.[3] In a cluster analysis, Bruehl et al[3] arrived at 3 relatively consistent CRPS patient subgroups that did not have notably different pain duration, suggesting the existence of 3 CRPS subtypes, not stages. Their study found that one of the subgroups best represented the clinical presentation of CRPS type II. This subgroup had the greatest pain and sensory abnormalities and the least vasomotor dysfunction of all 3 subgroups. Nonetheless, this study has not settled the discussion, as it only included 113 patients.[3] Thus, with future studies, our understanding of CRPS in stages may change, which likely will impact how the clinical diagnosis is made.

There is a lack of high-quality evidence for most treatment interventions for CRPS[8]; however, the current practice is to use an interdisciplinary approach.[1,9,10] The main therapeutic arm of this approach is rehabilitation; physical and occupational therapy can help improve range of motion, contracture, and atrophy. The other 2 arms of the approach are psychologic therapy to improve quality of life and pain management with pharmacologic therapy and/or invasive interventions. The choice of therapy remains empirical; trial and error should be expected in developing an adequate treatment plan for each individual patient.

Many aspects of CRPS remain unclear, and even our current understanding of the disease will inevitably change over time. The syndrome can cause life-changing morbidities in patients, and late diagnosis and treatment are associated with poor prognosis. Because there are many dermatologic findings associated with the disorder, it is crucial for dermatologists to clinically recognize the disorder and to refer patients to appropriate channels so that treatment can be started as soon as possible.

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