Case Report/Case Series

Dermatological Findings in Early Detection of Complex Regional Pain Syndrome

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**IMPORTANCE** Complex regional pain syndrome (CRPS) is a chronic pain condition usually affecting the extremities. It mostly occurs in 3 distinct stages with intense pain being the hallmark feature in every stage. Skin abnormalities are common, and often necessary, in the clinical findings required to diagnose CRPS.

**OBSERVATIONS** A man in his 30s presented to the dermatology clinic with complaints of recurrent redness, swelling, and burning pain in his left arm. Based on this clinical presentation with normal findings from a neurological examination and unremarkable findings on diagnostic imaging, the diagnosis of CRPS was made.

**CONCLUSIONS AND RELEVANCE** It is important for dermatologists to understand and recognize CRPS as a neurological disorder with major dermatologic implications. The ability of dermatologists to identify and direct patients with this syndrome is a critical factor in determining the likelihood of favorable outcomes following diagnosis of CRPS. This report outlines and reviews a neurological condition presenting with clinically significant cutaneous changes. We illustrate the bias that dermatologists may have in exclusively associating patient complaints with dermatological implications. This stresses the necessity for dermatologists to perform comprehensive medical histories and physical examinations to minimize diagnostic error and improve patient care.

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**ARTICLE INFORMATION**

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Comprehensive regional pain syndrome (CRPS) is a chronic pain condition usually affecting the extremities. The most common form of CRPS is type 1 (CRPS 1) and was previously referred to as reflex sympathetic dystrophy. The hallmark feature of CRPS is intense pain that gets progressively worse over time. Additional characteristics include impaired motor function, localized redness and swelling, pain produced by harmless stimulation (also known as allodynia) and joint stiffness. Complex regional pain syndrome typically exists as a triphasic disorder with an early stage described as pain in a limb, with warm extremities that progressively cool followed by atrophy as the condition worsens. Skin abnormalities usually increase in severity with later stages of the syndrome. No definable causes for CRPS have been agreed on, but patients commonly present with signs and symptoms following peripheral trauma, surgery, or vascular events, such as stroke and myocardial infarction.

Herein, we present the case of a patient with chronic pain and redness of the left arm that can be attributed to type 1 CRPS.

Report of a Case

A man in his 30s complained of a year-long problem of recurrent redness, swelling, and burning sensation developing on his left arm. It occurred 20 to 30 times daily (Figure). He also described ongoing pain that was out of proportion if he was touched on his left elbow, wrists, and possibly left knee, with associated numbness and motor dysfunction of the hand. The patient had no family history of multiple sclerosis or any other neurological conditions. He did not correlate his symptoms with any relatable inciting events but did recall an uncomplicated minor trauma to his scalp 2 months prior to the onset of symptoms when an overhead door hit his head and possibly his left elbow. Initially, the patient had a normal findings from a skin examination. Ten minutes into the interview, he showed clear evidence of gradual, mild erythema, as well as swelling to the left forearm with an increased temperature to the area. A previous magnetic resonance image of the head showed no evidence of radiculopathy despite ongoing sensory loss in his left palm. Findings from a prior neurological examination and electromyography were both interpreted as normal by the neurological department. This patient’s clinical history and presentation may, in part, be explained by conditions such as thoracic outlet syndrome, multiple sclerosis, or other neurological diseases. However, these possibilities were ruled out after the patient’s initial workup and were not explored in any further detail. Therefore, based on these observations, the patient was diagnosed as having CRPS with the erythema presenting secondary to neurologic stimulation. The patient was referred to...
Complex regional pain syndrome presents as a constellation of nonspecific symptoms and thus is diagnosed clinically. However, diagnostic testing can provide the clinician with more confidence in the diagnosis and help eliminate other potential disorders related to the associated symptoms of CRPS. Autonomic testing has been shown to predict the diagnosis of CRPS with good reliability, but the results are not unique to CRPS. Autonomic testing is performed by measuring a patient’s autonomic response to specific stimuli. Autonomic variables, such as blood pressure, heart rate, and skin temperature, are measured, and the response to stimuli is recorded and interpreted. Imaging studies that may be useful in the investigation of CRPS include bone scintigraphy, plain film radiography, and magnetic resonance imaging. Preventative measures, such as early mobilization after injury and ascorbic acid administration following fracture, have been suggested in reducing the risk of developing CRPS. Once a patient is diagnosed as having CRPS, treatment is supportive. Pharmacologic therapy has shown to be effective in pain relief and management of other symptoms of CRPS. Anticonvulsants, bisphosphonates, glucocorticoids, and calcitonin have shown to be more effective in pain management when compared with placebo. Sympathetic blocks with antiarrhythmic agents and local anesthetics are examples of invasive pharmacological interventions that have some efficacy in CRPS-associated pain management. The prognosis of CRPS is quite good if treatment is started early. A population-based study illustrated that 74% of patients reported resolution of symptoms. Nonetheless, without adequate therapy, CRPS can spread to other limbs and worsen in terms of symptoms and chance of recovery.

Skin changes are common and often-critical features required for the diagnosis of CRPS. Results from a retrospective study have shown that 74% of patients reported resolution of symptoms. Nonetheless, without adequate therapy, CRPS can spread to other limbs and worsen in terms of symptoms and chance of recovery.
medical chart review determined that cutaneous manifestations of vascular origin were most prominent in patients with CRPS. Edema and erythema were found in most patients, while dermatitis, erythematous papules, folliculitis, cutaneous atrophy, ulceration, and bullae were also seen to a lesser degree among the individuals in the study.\textsuperscript{13} The nonspecific nature of the associated skin findings makes it difficult to associate them with this syndrome because these patients are often misdiagnosed as having conditions that are more relevant to dermatology. Complex regional pain syndrome may be confused with a number of conditions with similar skin findings.\textsuperscript{14} Some of these include erythema migrans, acrodermatitis atrophicans, lipodermatosclerosis in chronic venous insufficiency, eosinophilic fasciitis, and scleroderma. Despite the prominent cutaneous abnormalities associated with CRPS, this condition has not gained much recognition among dermatologists. Araki et al\textsuperscript{15} reported that of 319 articles that contained “reflex sympathetic dystrophy”, only 1 was found in dermatology literature. This may indicate the failure of dermatologists to diagnose CRPS and suggest a necessary improvement in the recognition of skin abnormalities originating from unlikely sources, such as neurologic stimulation.

**Conclusions**

The early identification and management of CRPS are key factors that determine the likelihood of favorable outcomes following diagnosis and treatment. Although patients with CRPS are most often treated by specialists in the fields of neurology and anesthesiology, the complex nature of this neurological condition requires physicians in multiple specialties, including dermatology, to work together and treat these individuals. The dermatologic manifestations of this syndrome results in patients being referred to dermatology clinics for a neurological disorder. This warrants dermatologists to appreciate CRPS and take comprehensive medical histories and perform physical examinations to minimize diagnostic error and ensure the highest quality of patient care.

**REFERENCES**