Complex Regional Pain Syndrome
Diagnosis and Treatment

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KEYWORDS

• Complex regional pain syndrome • Neuropathic pain syndromes
• Reflex sympathetic dystrophy sympathetic nerve block • Spinal cord stimulation

KEY POINTS

- Complex regional pain syndrome (CRPS) is characterized by pain out of proportion to the usual time or degree of a specific lesion.
- The diagnosis of CRPS is based on 4 distinct subgroups of signs and symptoms: sensory, vasomotor, sudomotor, and motor/trophic changes.
- Treatment should be multidisciplinary, consisting of medications, physical/occupational therapy, psychotherapy, and sympathetic blocks targeted toward pain relief and functional restoration.
- More aggressive treatment, such as sympathectomy and spinal cord stimulation, have a low level of evidence but may be considered for therapy-resistant CRPS type I.

INTRODUCTION

Complex regional pain syndrome (CRPS) is characterized by pain that is out of proportion to the usual time or degree of a specific lesion. It does not present within the distribution of one peripheral nerve or nerve root, and has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor, and/or trophic findings. Progression is variable.[1] CRPS has been known by many other names including reflex sympathetic dystrophy (RSD) and causalgia. These terms date back to Claude Bernard, who in 1851 referred to a pain syndrome that was accompanied by changes in the sympathetic nervous system. During the American Civil War, Silas Weir Mitchell described cases of soldiers suffering from ongoing burning pain after recovering from gunshot wounds, and coined the term Causalgia.[2,3] Evans first used the term reflex sympathetic dystrophy in the 1940s to emphasize that the sympathetic nervous system

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was involved in the pathophysiology of the disease.[4] CRPS replaced the term RSD for several reasons. Sympathetic changes and dystrophy may not be present throughout the disease course.[5,6] Furthermore, there is no specific reflex arc that is responsible for the CRPS; pain is secondary to multisynaptic pathologic changes involving the brain, spinal cord, and peripheral nerves.

EPIDEMIOLOGY

CRPS has a female to male ratio of 2:1 to 4:1, which is more common with increasing age. There are 50,000 new cases of CRPS in the United States annually. The most common initiating events of the syndrome include fractures, sprains, and trauma such as crush injuries and surgery. Immobilization after injury is a contributing factor in more than half of patients.[7,8]

DIAGNOSIS

The Budapest Consensus Workshop introduced criteria to identify patients with CRPS and exclude other neuropathic conditions. More stringent criteria are used for research purposes to eliminate false-positive inclusions. Less stringent criteria are used in the clinical setting to avoid missing the diagnosis. A patient must report symptoms of, and display signs on physical examination, in the following categories: sensory, vasomotor, sudomotor/edema, and motor/trophic (**Figs. 1** and **2**). For both clinical and research purposes, a patient with CRPS should have physical examination evidence of at least 1 sign in 2 or more of the categories. The symptom criteria are different when assessing patients in a clinical rather than a research setting. In a clinical setting, patients must report 1 symptom in 3 out of the 4 categories, whereas in the research setting the patient must report 1 symptom in each of the 4 categories (**Box 1**). This minor adjustment in data collection creates a sensitivity of 0.85 and a specificity of 0.69 for the research group, compared with the clinical criteria that have a sensitivity of 0.94 and a specificity of 0.36.[1,9]

There are 2 subgroupings of CRPS. CRPS I is CRPS without major nerve damage (formerly known as RSD) while CRPS II is CRPS with major nerve damage (formerly known as causalgia). A third subtype is CRPS NOS (not otherwise specified), which captures patients who only partially meet the current criteria but were diagnosed with CRPS under previous criteria.[1,6,9]

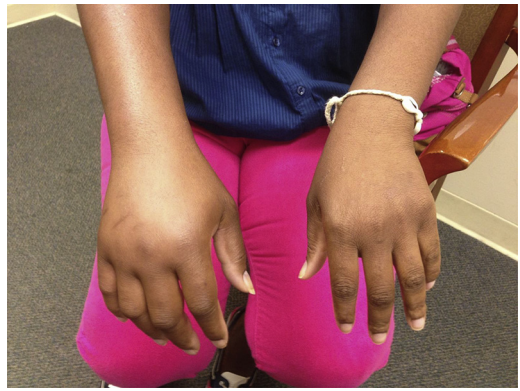

Fig. 1. A patient with 3 months of pain following brachial plexus injury has significant fusiform edema and color changes in the right upper extremity.