Research Paper Review

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Complex Regional Pain Syndrome

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Background Information
Complex regional pain syndrome (CRPS) – known historically as causalgia, reflex sympathetic dystrophy, Sudeck's atrophy, algoneurodystrophy, or shoulder-hand syndrome – is a chronic condition characterized by constant regional neuropathic pain that is usually associated with abnormal sensory, autonomic, motor and/or trophic changes (1). It generally develops after trauma, although it can also be idiopathic in origin. CRPS presents as one of two subtypes: type I (where no overt nerve lesion can be identified) and type II (where a definite nerve lesion is evident).

The pathophysiology of CRPS is multifaceted and not well understood. Several mechanisms have been suggested, including abnormal response to tissue injury, peripheral and central pain sensitization processes, neurogenic inflammation, endothelial dysfunction, disturbed sympathetic-afferent coupling, hyperalgesic priming, somatosensory cortical re-organization, genetic predisposition, and even some degree of autoimmunity (2), although no definitive explanation has been agreed upon.

The purpose of this paper was to provide a clinical review of the current literature and provide information on epidemiology, diagnosis and management of CRPS.

SUMMARY

Epidemiology
The true incidence of CRPS is uncertain. Only 2 retrospective studies have been completed on CRPS, with one finding an incidence of 5.46 cases per 100,000 person-years and a prevalence of 20.57 cases per 100,000 people (3), and a second estimating the incidence at 26.2 cases per 100,000 person-years (4). In general, female:male ratio ranges from 3.4-4:1, with a peak incidence in patients aged 50-70.

More recent studies investigating the incidence of CRPS following trauma found that CRPS occurred in 3.8% of 1549 patients within 4 months of a wrist fracture (5) and in 7% of 596 patients within one year of wrist/scaphoid/ankle/metatarsal fracture (6).

Clinical Features

Precipitating factors for the development of CRPS include fracture, soft-tissue injury or surgical trauma, although it can also be triggered by other conditions such as prolonged immobilization, disorders of the central nervous system (such as stroke), and visceral lesions (such as myocardial infarction). Idiopathic onset is estimated in 10% of cases.

The primary difficulty in diagnosing CRPS is the diversity of symptoms, both between patients and within the same individual. Key clinical features of CRPS, which tend to persist beyond the expected timeline associated with the initial injury, include:

- spontaneous pain,
- vasomotor changes,
- motor abnormalities,
- hyperalgesia (exaggerated pain to a painful stimulus such as pinprick), and/or
- allodynia (pain elicited by a normally non-painful stimulus such as light touch).

These symptoms can present mildly in the initial stages of CRPS and may evolve or change during the course of the condition, adding to the confusion and difficulty for the clinician. While some have suggested adoption of a three-stage concept to describe the progression of CRPS, there is insufficient evidence to support this as a natural course of the condition and it is therefore considered invalid (7).

Both neuropathic and nociceptive pain can coexist in areas affected by CRPS, although clinicians should distinguish between nociceptive patterns (sharp, achy, throbbing) and the neuropathic patterns associated with CRPS (burning, tingling, stabbing, numbness, electric 'shock'). Vasomotor and sudomotor changes also accompany the neuropathic pain, presenting as changes in temperature, skin colour, moistness, or edema (8). These changes are variable and can evolve over time. The changes can include:

- warmth and pink colour (or cold, brownish and mottled) and/or cyanosis in the affected region,
- atrophic and shiny skin,
- asymmetry or changes (increases or decreases) in sweating and in hair and nail growth, as compared with the unaffected side,
- hypertrophic or atrophic nail growth, with nails appearing brittle and ridged,
- abnormal motor function such as weakness, difficulty in initiating movements, tremor, muscle spasms, and dystonia, and
- disturbances in body perception including neglect and distorted mental image of the affected body part.

Natural Course of CRPS

Insufficient information is available to determine a consistent course of CRPS. Data from a 2009 study (9) indicated that the average time from the triggering event was 5.8 years and, at that point, 30% of patients considered themselves completely recovered, 16% reported severe symptoms and 54% were considered stable. Three factors were found to contribute to a worse outcome: having an upper extremity affected; having a triggering event other than a fracture; and being diagnosed with 'cold' CRPS, which is defined as CRPS associated with a self-reported 'cold' skin temperature up to 2 years following diagnosis.

Diagnosis

There are no validated tests specific for CRPS. Various tests including blood tests, plain films, nerve conduction studies and EMG studies have not demonstrated sufficient specificity or sensitivity. The

current recommendations for diagnosis involve the Budapest criteria (10), where each of the following criteria must be met for a diagnosis of CRPS:

- Patients must report continuing pain that is disproportionate in time and/or degree to the usual course of pain after any trauma or other inciting event,
- Patients must report at least one symptom in three of the four following categories:
    1. Sensory: hyperalgesia (that is, exaggerated pain to a painful stimulus, such as pinprick) and/or allodynia (that is, pain elicited by a normally non-painful stimulus, such as light touch).
    2. Vasomotor: skin colour and/or temperature changes/asymmetry.
    3. Sudomotor/edema: swelling and/or sweating changes or asymmetry
    4. Motor/trophic: weakness, tremor, dystonia, decreased range of motion and/or trophic changes/asymmetry involving nails, skin and/or hair.
- Patients must display at least one sign at the time of assessment in two or more of the same four categories listed above,
- Signs and symptoms must not be better explained by another diagnosis.

Treatment

A Cochrane review of systematic reviews on CRPS found that the current evidence for most proposed treatments is of low to very low quality and cannot be regarded as reliable (11). Treatments designed to address neuropathic pain, such as tricyclic antidepressants, serotonin-norepinephrine reuptake inhibitors, anticonvulsants, and topical lidocaine are commonly prescribed, with varying success. Bisphosphonates, used most often to prevent or slow bone loss in osteoporosis, have been recommended, but their use in CRPS is not supported by sufficient clinical evidence. There is currently no evidence to support conservative and/or manual therapies as a potential treatment to address the essential symptoms of CRPS (12). Rehabilitation practitioners can, however, make valuable contributions to the treatment of CRPS by providing education regarding management of chronic pain such as pacing, goal setting and relaxation. Multidisciplinary pain management programs focusing on functional restoration also have potential to provide relief from the symptoms of this difficult condition.

CLINICAL APPLICATION & CONCLUSIONS

CRPS is a debilitating condition associated with traumatic injury and one that is not associated with a reliable treatment option. While conservative treatment modalities may not provide consistent relief from symptoms, a focus on chronic pain management techniques may help to decrease the symptomatology. Clinicians should be aware of the presentation of CRPS and the appropriate co-management strategies that are likely to elicit the best results.

METHODS

A search of Medline, the Cochrane Library, and the website of the International Association for the Study of Pain (www.iasp-pain.org) using the terms “complex regional pain syndrome”, “causalgia”, “reflex sympathetic dystrophy”, “Sudeck’s atrophy”, “algodystrophy”, “algoneurodystrophy”, and “reflex neurovascular dystrophy” was conducted to identify potentially eligible papers. Articles relating to epidemiology, pathophysiology, natural course, diagnosis, and treatment were the primary aim of the search.

Additional References:

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