Complex Regional Pain Syndrome

Background

CRPS type 1 – Previously known as Reflex Sympathetic Dystrophy (RSD). "A syndrome of sustained burning pain, allodynia, and hyperpathia after a traumatic nerve lesion, often combined with vasomotor and sudomotor dysfunction and later trophic changes[1]." First defined by American Civil War physician, Silas Weir Mitchell, in his book titled *Injuries of Nerves and Their Consequences*[2]. Staging of RSD first proposed by John Bonica[4].

Stage 1 (acute) – From time of trauma up to 3 months after. Characterized by hyperalgesia/allodynia, vasomotor/sudomotor dysfunction, and edema.

Stage 2 (dystrophic) – Occurring 3 – 6 months after onset. Characterized by severe pain, sensory dysfunction, vasomotor dysfunction, and motor/trophic changes.

Stage 3 (atrophic) – Occurring after 6 months from onset. Characterized by decreased pain, increased sensory/vasomotor disturbance, and increased motor/trophic changes.

CRPS type 2 – Previously known as Causalgia. Originally described by James A. Evans as a chronic pain syndrome with signs of sympathetic stimulation[3]. Now referred to as CRPS Type 2. Typically occurs after soft tissue or bone injury.

International Association for the Study of Pain (IASP) 1994 Criteria[5]
1. The presence of an initiating noxious event, or cause of immobilization
2. Continuing pain, allodynia, or hyperalgesia in which the pain is disproportionate to any known inciting event
3. Evidence at some point of edema, changes in skin and blood flow, or abnormal sudomotor activity in the region of pain (can be sign or symptom)
4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction

The IASP 1994 criteria has good sensitivity but poor specificity which may lead to over diagnosis[6].

The Budapest Criteria[7]
1. Presence of continued disproportional pain from the known inciting event
2. Must report at least one symptom in three of the following four categories:
a. Sensory: reports of hyperesthesia, allodynia
b. Vasomotor: reports of temperature asymmetry, changes in skin color
c. Sudomotor/edema: reports of edema, changes in sweating, sweating asymmetry
d. Motor/trophic: decreased range of motion, motor dysfunction, trophic changes
3. Must report at least one sign in two or more of the following categories at the time of evaluation:
a. Sensory: evidence of hyperalgesia to pinprick, allodynia to touch or joint movement
b. Vasomotor: evidence of temperature asymmetry, color asymmetry
c. Sudomotor/edema: evidence of edema, asymmetrical sweating, sweat changes
d. Motor/trophic: evidence of decreased range of motion, motor dysfunction, trophic changes
4. There is no other diagnosis or condition that can better explain the signs and symptoms

The Budapest Criteria of 2003 has good sensitivity and improved specificity[6] and provided the distinction between CRPS 1 and CRPS 2

Treatment

The primary goal in the treatment of CRPS is restoration of function and alleviation of pain. A multidisciplinary treatment plan includes: psychotherapy, physical rehabilitation, pharmacotherapy, and interventional therapy.

Pharmacotherapy

- Membrane Stabilizers – Gabapentin, Pregabalin. First-line treatment medication for neuropathic pain.

- Antidepressants – TCA's (amitriptyline). SNRI's (venlafaxine, duloxetine). TCA's have shown to be more effective at treating neuropathic pain than SNRI's[8].

- Anti-Inflammatory Drugs – NSAIDs for the treatment of CRPS has mixed results and shows no benefit in CRPS type 1[9]. Oral corticosteroids have shown effectiveness in the treatment of CRPS[10].

- Opioids – Use in CRPS is controversial as long-term opioid use may exacerbate allodynia. Tramadol may be an effective treatment due to its SRNI attribute[7].

- Bisphosphates – Alendronate. May decrease pain in CRPS by slowing bone resorption during bone remodeling.

- Baclophen – Effective treatment for dystonia related to CRPS type 1[11].

- NMDA Receptor Antagonists – Ketamine, dextromethorphan. Ketamine infusion over 4 hours for 10 days has proved to be an effective treatment regiment[12].

- IV immunoglobin (IVIG) – research suggests that the immune system may contribute to sustained-CRPS. Intravenous immunoglobin has demonstrated the ability to reduce pain in some patients with CRPS[13].

Interventional Therapies

- Sympathetic Nerve Blocks – Stellate ganglion block (upper extremity involvement). Lumbar sympathetic chain block (lower extremity involvement).

- Epidural Infusion – Epidural infusions have shown beneficial while intrathecal analgesia infusions have proved ineffective[14]. A major drawback to the use of epidural infusions for the treatment of CRPS is the high incidence of infections related to this technique if the catheter remains in place for greater than 2 weeks[15].

- Spinal Cord Stimulation – Treatment option for late stages of CRPS

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