Complex regional pain syndrome type I

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The International Association for the Study of Pain has recently introduced a new title "complex regional pain syndrome" to replace the confusing terminology of reflex sympathetic dystrophy and causalgia. This review article highlights the diagnostic criteria used and illustrates the necessity for such a change. The role of the sympathetic nervous system and the efficacy of sympathectomy in patients with sympathetically-maintained pain are discussed. Controversies still exist regarding the use of various investigation tools, the results of which should be interpreted with caution. Finally, a multidisciplinary approach with different modalites to achieve the best success in breaking the cycle of pain and returning the patient to normal and productive function is the main aim of the treatment.

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Introduction

The terms causalgia, reflex sympathetic dystrophy (RSD) and its synonyms (Table 1) have caused much confusion within the medical profession. None of these terms are satisfactory in describing the underlying diseases, as no one understands the pathophysiology or knows the best treatment for them. In 1994, the International Association for the Study of Pain (IASP) agreed to introduce the title "complex regional pain syndrome" (CRPS) to replace these terms.[1] The old terminology of RSD is now termed CRPS type I, and causalgia is described as CRPS type II.

History

The word causalgia was coined by Silas Weir Mitchell in 1816 during the American Civil War to describe the burning pain that followed penetrating nerve injuries by bullets.[2] In 1900, Sudeck published the first classical description of the painful post-traumatic disorder associated with vasomotor disturbances.[3] Subsequently, many different terms have been used to describe similar disorders and the term RSD was used by Evans in 1946.[4] This merely reflects a lack of agreement among the medical profession on the possible aetiology and treatment of the underlying disease.

International Association for the Study of Pain diagnostic criteria

According to the IASP classification, there are four diagnostic criteria for CRPS type I:

1. The presence of an initiating noxious event, or a cause of immobilisation.
2. Continuing pain, allodynia, or hyperalgesia with the pain being disproportionate to any inciting event.
3. Evidence at some time of oedema, changes in skin blood flow, or abnormal sudomotor activity in the region of the pain.
4. The diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

Criteria 2 to 4 must be present to make the diagnosis of CRPS type I.

The diagnosis of CRPS type II requires the presence of the following three criteria:

1. The presence of continuing pain, allodynia, or hyperalgesia after a nerve injury, not necessarily limited to the distribution of the injured nerve.

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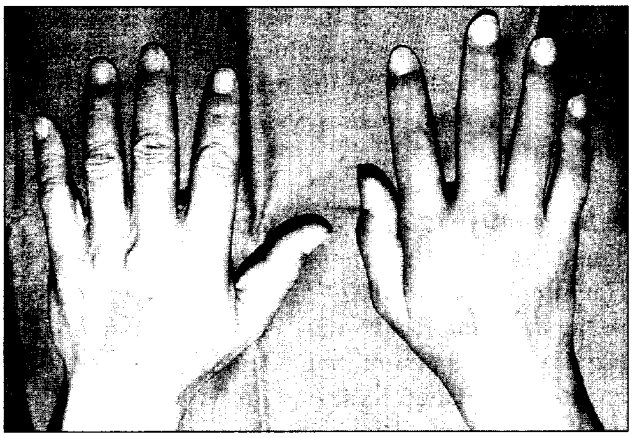

Figure 1. Hands of a patient with type I CRPS six months after injury. Note wasting of the forearm with ulnar deviation, dilated veins, oedema, and tapering of the fingers.

2. Evidence at some time of oedema, changes in skin blood flow, or abnormal sudomotor activity in the region of the pain.
3. The diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.

Physical symptoms and signs of the CRPS type I are not usually limited to the distribution of a single peripheral nerve, and the intensity is apparently disproportionate to the inciting event. It usually occurs at the distal aspect of an affected extremity or with a distal to proximal gradient. However, CRPS type II usually occurs in the region of the limb innervated by a damaged nerve after partial injury of the nerve or one of its major branches.

The role of the sympathetic nervous system

One advantage of the IASP classification is that the sympathetic nervous system is no longer directly implied in the diagnosis of the syndrome. The presence of vasomotor and sudomotor changes had previously suggested that sympathetic hyperactivity is the cause of the problem. In clinical practice, however, many patients fail to respond to appropriately conducted sympathetic blockades. Indeed, the sympathetic system can play either an active or a passive role—active, if there is a lesion within the sympathetic system and passive, if the sympathetic system is activated in response to a primary insult. Sympathetic blockade often does not eliminate the pain associated with the latter mechanism. The terms "sympathetically dependent pain" or "sympathetically maintained pain" have been used to describe a subset of patients in whom pain relief and reversal of associated sensory disorders can be achieved by appropriate blockade of the sympathetic nervous system.[5] In contrast, the term "sympathetically independent pain" has been used for patients with clinical features of CRPS type I but who fail to respond to appropriate sympathetic blockade.

Table 1. Terminology used to describe reflex sympathetic dystrophy

Acute bone atrophy
Algoneurodystrophy
Causalgia - minor, major, minimo
Chronic traumatic oedema
Lechirche's post-traumatic pain syndrome
Minor traumatic dystrophy
Post-traumatic pain syndrome
Post-traumatic painful osteoporosis
Post-traumatic spreading neuralgia
Post-traumatic vasomotor disorders
Reflex neurovascular dystrophy
Shoulder-hand syndrome
Sudeck's atrophy
Sympathalgia
Sympathetic maintained pain
Traumatic vasospasm

Neuropathic pain

Neuropathic pain is an important feature of the CRPS type I. It is frequently described as a continuous burning sensation and is often exacerbated by movement, continuous stimulation, or stress. Allodynia or hyperalgesia may be present and is not restricted to the territory of a single peripheral nerve. This led to the belief that the primary lesion is within the nervous system and Woolf suggested that this may represent a maladaptive neuronal plasticity involving the peripheral and central nervous system.[6] Not all nerve injuries, however, result in pain. Because long term changes in spinal plasticity occur only in a small proportion of patients with nerve injury, it has been suggested that this may represent a random error within the nervous system.[7] If one examines the results of treatments performed for this condition, there are no reliable predictors of successful outcome. Therefore, it seems that the treatment and the result of the therapy are random events also.

As the disease progresses, the picture is complicated by the presence of nociceptive pain secondary to tissue disuse, pseudo-paralysis, and contractures[8] (Figure 1). This may account for the observation that sympathetic blockade alone does not provide adequate

analgesia in patients with long term symptoms. Many patients in the late stage of the disease develop psychological disturbances, including anxiety and depression. These are often the result, rather than the cause of prolonged pain and disability.[9]

Diagnostic considerations

The last criterion for CRPS type I requires the exclusion of correctable causes such as minute fractures, connective tissue diseases, and vasculopathy. There is no single laboratory test for confirming the diagnosis of CRPS type I. Local anaesthetic sympathetic ganglion blockade has long been used as the main diagnostic and therapeutic tool. This is especially true when surgical intervention such as sympathectomy is contemplated. In cases with sympathetically-maintained pain, sympatholytic intervention may provide temporary or permanent pain relief.

Local anaesthetic technique

Cervico-thoracic stellate and lumbar sympathetic ganglia can be blocked with percutaneous injections of long acting local anaesthetic agents such as bupivacaine. The result of the blockade must be interpreted with caution as false positive results can occur in the following circumstances:

1. Presence of placebo response, which may occur in 30% to 40% of patients.[10]
2. During cervico-thoracic sympathetic block, local anaesthetic can diffuse posterolaterally to the cervical nerve roots resulting in subtle somatic nerve fibre blockade that can be difficult to detect clinically but may affect the patient's pain.
3. The systemic absorption of local anaesthetic has been shown to produce pain relief in various conditions of neuropathic and central pain syndromes.[11] Mexiletine, an oral form of lignocaine, has been used successfully in treating some patients with painful diabetic neuropathy.[12]

There is always the chance of obtaining a false negative result if the block is inappropriately performed. A well conducted local anaesthetic sympathetic blockade should be performed by an experienced doctor who should carefully examine the sensory system of the patient before and after the procedure. The degree of pain relief should be documented over time and the adequacy of sympathetic blockade should be monitored. For instance, the presence of Horner's syndrome, a rise in skin temperature, or abolition of the vascular constrictor reflex in response to deep inspiration on the side of the blocked extremity[13] indicate a successful cervicothoracic sympathetic block. Finally, the result of the test should be correlated with the following types of sympathetic blocks.

Intravenous regional technique

This less invasive technique was first proposed by Hannington-Kiff in 1974[14] and is commonly quoted in the anaesthetic literature. This technique makes use of the sympathomimetic depletion properties of agents like guanethidine and bretylium. The procedure is similar to Bier's blocks performed for operations on the upper or lower extremities. Guanethidine depletes the intravesicular noradrenaline storage in the presynaptic nerve endings. Pain relief may last for two to three days in patients with sympathetically-maintained pain. However, recent randomised, prospective controlled studies, have not shown any significant advantage in using guanethidine over placebo in terms of pain relief and long term outcome.[15,16]

Intravenous phentolamine test

Phentolamine is a short acting non-specific alpha antagonist. It can be infused slowly via a peripheral vein to produce sympathetic blockade.[17] The procedure is easy to perform and causes minimal discomfort to the patient. It also allows for a placebo trial to assess the variability of the patient's pain response. However, a dose-response curve is not obtained. The recommended dose varies from 0.5 to 1.0 mg/kg. Abolition of the reflex vasoconstrictor response may be used as an endpoint for assessing the completeness of the sympathetic block.

Dynamic bone scanning

Three-phase bone scanning has gained popularity in recent years for the diagnosis of RSD. An analysis of the literature shows a wide variability in scintigraphic accuracy in patients with clinically obvious RSD.[18] Bone scintigraphy is preferred to radiography for the early diagnosis of post-fracture type I CRPS and the technique of delayed rather than three-phase bone scanning seems sufficient. Other than this, there is no advantage of bone scanning over radiography in diagnosing late stages of the disease.[19]

Thermography

The usefulness of thermography remains controversial, as temperature changes of the extremities are influenced by many factors.[20] Thermography provides information about skin temperature but is not diagnostic of any disease state.

Management

A detailed description of the management of these syndromes is beyond the scope of this discussion. Readers are directed to other review articles which all agree to disagree on what is the correct therapy.[21,22] It is important to tailor the treatment individually. The main aims of the therapy are to restore normal function and ameliorate the suffering from pain. Best results will be achieved if the treatment is started early and adapted to the clinical stage of the disease.[23] Sympathetic interruption,[24,25] neural blockade,[26] corticosteroids,[27] calcitonin,[28] beta-blocking agents, and more recently, biphosphonates, have been advocated. As the disease progresses, treatment becomes more difficult and an adverse outcome is more likely.[29] The pain can recur, even after amputation of the severely affected extremity.[30]

The strategies of management are as follows:

1. Identify and correct any precipitating cause as far as possible.
2. Determine whether there is any sympathetic component. If so, consider chemical or surgical sympathectomy.
3. Functional rehabilitation with aggressive, active or passive physiotherapy. There is evidence that early disease may respond to exercise alone.[31] On the contrary, pain relief without exercise is unlikely to be successful. Aggressive physiotherapy and occupational therapy form the cornerstone of treatment.
4. Treatment for late disease is especially difficult when nociceptive pain secondary to tissue disuse and contractures sets in. Conventional therapy for nociceptive pain, including the use of opioids, may have to be used. Recently, both intraspinal opioids[32] and dorsal column stimulators[33] have seemed to produce favourable results in terms of pain relief.
5. Emotional aspects need to be addressed.[34] The pain and disability is so distressing to patients and their families that suicide is not unheard of. It is important to explain to the patient that it is normal to have an emotional response to the pain and also to the disability. When this response interferes with successful physical therapy, however, it must be addressed.

Conclusion

Type I CRPS remains largely a clinical diagnosis and depends very much on the exclusion of correctable causes. There is no definitive laboratory test. Early recognition and aggressive physiotherapy with adequate pain relief is important. However, long-term outcome remains unsatisfactory especially when treatment is delayed. Sympathectomy may be helpful in a subgroup of patients only. A multidisciplinary approach with all available modalites to achieve the best results in breaking the cycle of pain and returning the patient to normal and productive function is the essence of the treatment. The IASP reclassification will enable us to have a more uniform diagnosis and, hopefully, a better assessment of the therapeutic options will result.

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