Complex regional pain syndrome (CRPS)

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CRPS is a debilitating, painful condition that usually affects a limb, the arm and hand more commonly than the lower extremity. It is of unknown aetiology. The term CRPS was introduced to combine a number of conditions, eg reflex sympathetic dystrophy, causalgia and Sudeck's atrophy, that are thought to share a common mechanism. CRPS is divided into Types 1 and 2; the latter term is used where there is clear evidence of nerve injury, although the general principles of management are the same.

As it is a syndrome, CRPS is diagnosed in terms of symptoms and signs; there are no investigations that will confirm or refute the diagnosis. The Budapest criteria (see Royal College of Physicians guidelines) are widely used for this purpose, with a positive diagnosis being dependent on either symptoms or signs in three or two respectively of four categories: sensory, vasomotor, sudomotor/oedema and motor/trophic. The patient should have persistent pain disproportionate to the triggering event and no other diagnosis can better explain the findings.

Transient CRPS is relatively common and may occur in up to 25% of patients after minor trauma but, fortunately, severe CRPS is relatively uncommon. About 15% of CRPS sufferers will have unrelenting pain with significant disability lasting more than five years. It is regarded as a random error within the nervous system in genetically-predisposed individuals, ie someone predisposed to the condition may have had many uncomplicated injuries prior to developing the condition. There is evidence of increased sensitivity in peripheral neurones to neuro-transmitters, including noradrenaline with, importantly, central sensitisation in the brain. Functional magnetic resonance imaging scans show altered activity in the contra-lateral motor and somato-sensory cortical areas and patients demonstrate laterality, ie they are slower and less accurate in identifying images of a hand of the affected side compared to the other side. Pre-existing psychological problems do not predispose to the development of CRPS, but may impact on management. In the author's experience, many patients developing CRPS complain of unsatisfactory management of their triggering injury with severe pain and an over-tight POP. It is likely that this is the result of developing of CRPS rather than being the cause.

TREATMENT

There is no medical cure for CRPS. The four pillars of treatment are patient education and reassurance, pain relief, physical and vocational rehabilitation, and psychological intervention.

Treatment of established CRPS requires a multidisciplinary approach, but management should begin in primary care with early diagnosis and use of conventional analgesics and, if necessary, neuropathic pain medications, eg amitriptyline and gabapentin (as per NICE guidelines for the management of neuropathic pain). The patient should be encouraged to perform gentle exercises with the affected limb (unless contra-indicated) with reassurance that 'hurt does not mean harm'.

If the condition does not show clear evidence of resolution, specialist help should be sought. Involvement of orthopaedic surgeons is important to exclude treatable underlying conditions and repeated assessments may be necessary. Physiotherapists and occupational therapists have a key role as restoration of function appears to be critical to a positive outcome. In addition to gentle active movements, techniques such as mirror-box therapy (to try to re-programme the brain) and desensitisation can be very helpful. Pain Medicine has a role in bringing the four pillars of treatment together, as well as managing more complex analgesic regimes including IV pamidronate and, possibly, spinal cord stimulation. Psychological interventions may be offered by specialist services, but all healthcare staff involved with patients with CRPS have a part to play in tackling general issues such as anxiety, depression and sleep disturbance as well as more specific problems such as addressing unhelpful beliefs and behaviours.

The diagnosis and management of CRPS was greatly enhanced by the publication in 2012 of evidence-based guidance by a multidisciplinary group via the website of the Royal College of Physicians. The guidelines include general information and speciality-specific advice. In addition, there is a section with patient information, which supports the efforts of healthcare staff. This can be accessed at:

<http://www.rcplondon.ac.uk/resources/complex-regional-pain-syndrome-guidelines>

I strongly advise anyone who may be confronted by a patient with what could be CRPS to study the six pages of the concise guidelines, even if you don't venture into the 84 pages of the full guide.

COMMENTARY

RCP guideline author, Dr Andreas Goebel, senior lecturer in Pain Medicine at the University of Liverpool, comments on research projects in Liverpool

At the Walton Centre in Liverpool, we run a specialised service for patients with longstanding and/or complicated CRPS. We receive four to five referrals of patients in this group per month. As one of the worldwide leading centres for this condition, we also conduct both clinical and laboratory research studies. We are leading in exploring the possible autoimmune genesis of CRPS, and consequently in exploring new ways of treating these patients using anti-autoimmune treatments. In addition, we investigate novel physiotherapy/occupational therapy, and neuromodulation techniques. A large randomised controlled trial on low-dose intravenous immunoglobulin treatment for this group (moderate or severe CRPS of >1 year duration) is expected to start this spring.